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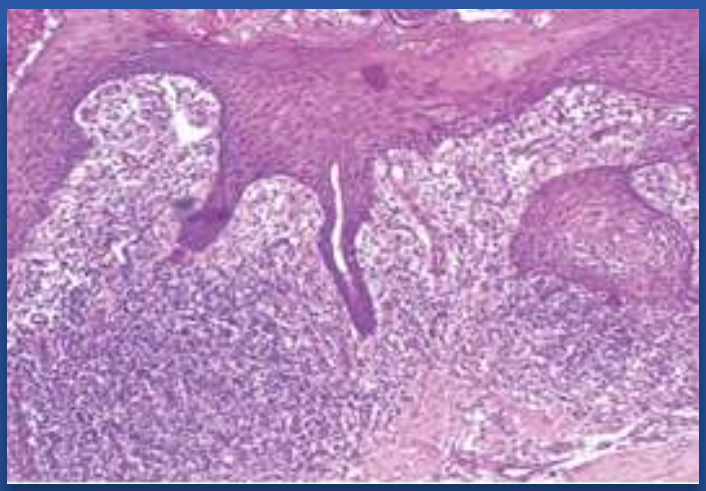

HIGHLIGHTS

Sunscreens: The Current Scenario

Trichoscopic Findings in Various Scalp Alopecias

Unilateral Truncal Acne After Laminectomy

Armoured Keloid

Werner's Syndrome: A Rare Entity

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CONTENTS

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FROM THE DESK OF EDITOR

Greetings to all the readers !!

I present to you another issue of Indian Journal of Clinical Dermatology. With your love and support we have entered second year of publication. We started with a goal to give space to maximum research publications in our journal so that they reach the people. However maintaining the quality of publications has also been taken care of.

We sincerely thank our authors for entrusting us with their valuable research work. Without their support this journey would not have been possible. I also thank our reviewers for sparing their precious time to evaluate our articles.

I hope you like our issue. I also request everyone to contribute towards journal with their valuable research work.

Dr. Dinesh Mathur
Editor

REVIEW
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SUNSCREENS: THE CURRENT SCENARIO

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Abstract

The increasing incidence of cutaneous malignancies and detrimental effects caused by ultraviolet radiation (UV) has increased the use of sunscreens. Many organic and inorganic filters are used as a measure of photoprotection, but their efficacy and safety profile still raise questions. Concerns have been raised regarding safety of nanotechnology in sunscreen, detrimental effects on environment, photocarcinogenic potential of UV filter to name a few. New developments in formulations of sunscreens along with changes in the guidelines of the regulatory bodies like The United States Food and Drug Administration and European Union have prompted us to revisit this topic. Continuous public education is still needed about proper application technique of sunscreen along with behavioral measures. In this article, the authors try to shed light on classification, pharmacological actions, various related terminologies, indications, emerging concerns and correct application technique as well as usefulness of oral sunscreens.

Key Words - Ultraviolet radiation, sunscreen, photoprotection, controversies.

Introduction

Photoprotection is crucial to prevent detrimental effects of ultraviolet (UV) radiations like photo-carcinogenesis, photoageing and photosensitivity. Sunscreens have become an integral part of not only the dermatologists' therapeutic armamentarium but are also distributed as over the counter and cosmetic products because of increasing awareness against harmful impact of radiations. Nevertheless, we need to continue educating the general population about photo protective measures as there are concerns of inadequate application of sunscreen. In this article we attempt to compile basic aspects of sunscreens along with an effort to understand debatable issues associated with it.

Why do we need photo protection?

Ultraviolet radiations (wavelength 200-400nm) are a small part of electromagnetic radiation spectrum, classified as UVA, UVB and UVC. Most detrimental and probably extensively studied part of UV radiations is its role as a major causal factor of skin cancer.[1]Unprotected chronic sun exposure leads to development of non melanoma skin cancers.[2] Direct photochemical damage to DNA is caused by UVB leading to gene mutations by means of pyrimidine dimers and development of precancerous and cancerous lesions while UVA penetrates deeper into skin; acting indirectly at cellular level by generation of free radical species.[3] Ultraviolet radiations cause both acute and chronic effects on skin which are elaborated in [Table 1].[4],[5]

Majority of current research and preventive strategies are centered around detrimental effects of UV radiations on skin due to its higher photon energy and relatively visible macroscopic changes. However, visible light which has been less studied so far, has a significant role in disease pathogenesis like solar urticaria, porphyria and idiopathic photodermatoses.[6] Infrared

Table 1: Effects of ultraviolet radiations on skin.

Ultraviolet radiation spectrum (200-400nm)			
	UVC	UVB	UVA
Wavelength (in nanometers)	200-290	290-320	UVA1=340-400 UVA2=320-340
Sea level solar radiation	0% (Completely absorbed by ozone)	Approximately 2-5%	95-98%
Molecular and cellular effects		• Cyclobutane pyrimidine dimer • 6-4 pyrimidine-pyrimidone dimer • Epidermal sunburn cell • Skin hyperplasia • Vitamin D synthesis	• Reactive oxygen species •Immuno-suppression •Cyclo butane pyrimidine dimer (weak)
Clinical effects Acute		• Erythema (peaks 24 hours), Oedema • Pigment darkening • Delayed tanning	• Immediate pigment darkening (fades within 15 minutes)
Chronic		• Photocarcinogenesis • Photoageing • Immunosuppression (weak)	• Photoageing •Immunosuppression • Photocarcinogenesis (weak)

A (IRA) can cause photoageing and photocarcinogenesis through its ability to induce gene alterations.[7] Education about wholesome photo protective measures is needed to attain overall protection against solar radiations.

Sunscreen usage: indications

Sunscreens have become an integral part of day to day activity, primarily used for protection against immediate and long term ill effects of ultraviolet radiations. With current trends of leisure activities like sunbathing, tan beddings and increased awareness of skin cancers, markets are flooded with more and more sunscreens. Sunscreens are primarily indicated in prevention and management of freckling, sunburn, photoageing, photocarcinogenesis, photosensitive and photo-aggravated dermatoses. Sunscreens have become indispensable in procedure driven dermatology to prevent post procedural hyper pigmentation. Strict photoprotection is needed to prevent development or aggravation of certain dermatoses, few examples of which are tabulated [Table 2].

Table 2. Indications of strict photoprotection.

- Dysplastic naevus syndrome
- Systemic lupus erythematosus
- Dermatomyositis
- Genetic skin cancer syndromes (xeroderma pigmentosum, Gorlin syndrome)
- Bloom syndrome, Cockayne syndrome
- Previous or current non-melanoma skin cancer
- Previous melanoma
- Previous exposure to arsenic or ionizing radiation
- Patients on systemic immunosuppressive therapy
- Porphyrias

Sunscreen: classification and characteristics

Sunscreen agents are broadly divided into topical and systemic. According to the Food and Drug Administration (FDA), topical sunscreens are classified as organic and inorganic, discarding the previously used terms like chemical and physical sunscreens [Figure 1]. An organic sunscreen agent is an active chemical which depending on their chemical characteristic absorbs UV radiation thereby moving into higher energy state from ground state. Depending on the fate of higher energy excited state, these are further divided into photo-stable, photo-unstable and photo-reactive.

- *Photo-stable sunscreen:*
  - » It returns to the ground state after dissipating its absorbed radiation to the environment as heat.
  - » Subsequently becomes capable of absorbing UVR again (recycles).
- Photo-unstable sunscreen:
  - » It degrades or undergoes conformational change after absorbing UV energy.
  - » It cannot enter in next cycle.
- *Photo-reactive sunscreen:*
  - » These agents produce free radicals by interactions of their excited state to surrounding biological molecules.
  - » They can exert unwanted biological effects.

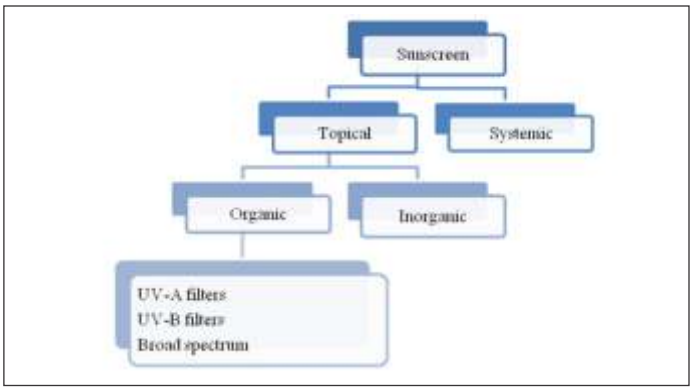

Figure 1. Classification of sunscreens.

Inorganic agents exert their protective function by means of reflecting, scattering or absorbing UV radiation. To enhance user's experience their "whitening effect" may be lessened by using their micronized or ultrafine particles.[8] Incorporation of inorganic particulates has an added advantage of scattering light from the upper layers of epidermis thus enhancing the sunburn protection factor (SPF) value.[9],[10]

On the basis of photo protective quality against particular wavelengths, topical sunscreens are classified as UVA filters, UVB filters and broad spectrum sunscreens. Sunscreen agents are recognized by three widely used nomenclatures which are the US adopted name (USAN), International Nomenclature Cosmetic Ingredient (INCI) name, and trade name [Table 3].[11]

Sunscreen related terminology:

For a long time the labeling information over sunscreens was misleading and confusing, making claims like sweat proof, water proof etcetera. Regulatory agencies like the FDA and European Union (EU) issued guidelines to regulate labeling of sunscreens hence enabling people to choose effective agents, to provide optimal sun protection without being misguided by lucrative claims of pharmaceuticals.[12] The FDA has discarded the terms like sweatproof, waterproof and sun blocks for the same reasons. These terminologies have been replaced by water resistant and very water resistant which are adequately defined. According to the FDA, manufacturers should display the effective duration of water resistance and need of reapplication over the sunscreen label. Water resistant sunscreens should be applied before activities like swimming or excessive sweaty conditions. False claims like immediate protection after application or longstanding efficacy of product (lasting >2 hours) are forbidden if not supported by enough evidence. Consumers can confidently choose a product suitable to their needs with help of this information over label and be realistic about photo protective efficacy of the product. The FDA has also made it mandatory to test a product for both UVB and UVA protection before using the terminology 'broad spectrum' on the label.

Water resistant- withstand 2 sequential water immersion of 20 minutes (40 minutes total) while maintaining claimed SPF value.

Very water resistant- maintains claimed SPF after immersion in water for 20 minutes, 4 times (80 minutes total).

Critical wavelength- wavelength at which 90% of the total area under the absorbance curve occurs.

Broad spectrum- sunscreen with critical wavelength ≥ 370nm with UVA protection factor ≥ 4.

Table 3. Classification and nomenclatures of topical sunscreen agents.

USAN	*INCI*	*Trade name*
Broad-spectrum and UVAI (340-400 nm)		
Bemotrizinol	Bis-ethyl hexyl oxyphenol mmethoxy phenyltriazine	Tinosorb S
Bisoctrizole	Methylene bis-benzotriazolyl tetramethylbutylphenol	Tinosorb M
Silatriazole	Drometrizole trisiloxane	Mexoryl XL
Ecamsule	Terephthalylidene dicamphorsulfonic acid	Mexoryl SX
Avobenzone	Butyl methoxydibenzoyl methane	Parsol 1789
	Diethylamino hydroxybenzoyl hexyl benzoate	Uvinul A Plus
Bisdisulizole	Disodium phenyl dibenzimidazole tetrasulfonate	Neo Heliopan AP
Zinc oxide	Zinc oxide	ZnO(nanox)
UVB (290-320 nm) and UVAII (320-340 nm)		
Enzacamene	4-Methylbenzylidene camphor	Eusolex 6300
Oxybenzone	Benzophenone-3	
Padimate O	Ethyl hexylrimethyl PABA	Eusolex 6007
Octinoxate	Ethyl hexylmethoxycinnamate	Uvinul MC 80
Octisalate	Ethyl hexyl salicylate	Neo heliopan OS
Amiloxate	Isoamyl p-methoxycinnamate	Neo heliopan E1000
Octyltriazone	Ethyl hexyltriazone	Uvinul T 150
Sulisobenzone	Benzophenone -4	Uvinul MS40
Octocrylene	Octocrylene	Uvinul N 539 T
Homosalate	Homomenthyl salicylate	Eusolex HMS

The efficacy of sunscreen agents is determined by two main indices; SPF and UV protection factor. There are several in vivo and in vitro methods to determine these. Protection against erythemogenic spectrum of UV light (UVB and UVA2) is measured by sunburn protection factor (SPF).[12]

The sunburn protection factor is measured as a ratio of the amount of UVR necessary to burn the protected skin (with sunscreen) to that required to burn the same non protected skin (without sunscreen) with all other parameters being constant. The required amount of UVR is known as MED (minimal erythema dose) which is defined as the minimum UV dose required to produce perceptible erythema of the skin with well defined margins at 16 to 24 hours after UV irradiation.[12] This means that a SPF 30 sunscreen protected skin can tolerate the same amount of UV radiation 30 times more than the unprotected skin. The grading of SPF is done as low (SPF 2-15), medium (SPF 15-30), high (SPF 30-50) and highest (SPF >50).

For measurement of UVA protection, various testing methods have been developed by regulatory bodies in Japan, the European Union (EU), United Kingdom (UK) and Australia.

UVA protection indices:

1. Australian/New Zealand Standard: In vitro method. 8-µm and 20-µm thick layers of the product should not transmit greater than 10% and 1% of radiation of 320 to 360 nm, respectively.
2. Japanese standard (persistent pigment darkening): In vivo test. It is a ratio of UVA required to induce persistent pigment darkening (PPD) 2 to 24 hrs after irradiation in sunscreen protected skin to unprotected skin. Ratings- PA+, PA++, PA+++, PA++++ (PA= protection grade from UVA).
3. European Union guidelines: Based on PPD method. It requires UVA protection factor to be ≥ 1/3 of labeled SPF.
4. Boots star rating system (United Kingdom): In vitro method. It measures ratio of UVA absorbance to mean UVB absorbance.

Since 2012, the FDA has mandated that only sunscreens having SPF≥15 and critical wavelength ≥370 nm, can display their claim about protection against development of skin cancers.[12]

Ideal sunscreen: The notion of ideal sunscreen is that it should provide maximum photo-protection while maintaining its compliance quality. An ideal sunscreen should possess 'spectral homeostasis', which refers to uniform protection against UVA

and UVB radiations spectrum.[13] The characteristics of an ideal sunscreen also include cosmetic acceptability and non-irritant nature, among others [Figure 2].

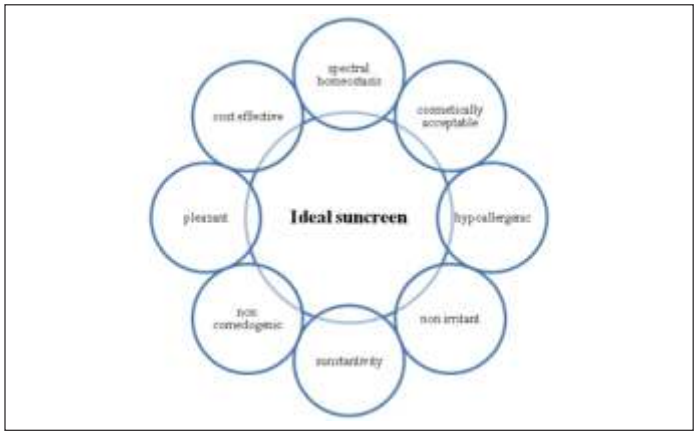

Figure 2. Ideal sunscreen

Guidelines for application:

Apart from SPF and substantivity, the other major factor that determines the protective efficacy of a given sunscreen is its adequate application. There is a lack of basic knowledge about correct application, leading to attainment of only one third to quarter of the recommended dose of photoprotection.[14] Sunscreen needs to be applied about half an hour before going outdoor, in a density of at least 2 mg per square centimeter which can be further simplified by following the "Tea spoon rule."[15] The rule recommends application of 1 teaspoon of sunscreen to the face/head/neck, 1 teaspoon to each upper limb, 2 teaspoons to trunk (front and back), and 2 teaspoons to each lower limb. The protective efficacy can be boosted significantly by reapplication 20 minutes later, thus correcting the areas of inadequate application.[6]

Sunscreen should always be used along with complete photoprotection package which includes seeking shade, protective clothing, wide-brimmed hat and sunglasses. A continuous effort in public education is required concerning its correct application, reapplication and prevention of unnecessary sun exposure.

Systemic sunscreens:

There is a growing interest in use of orally active ingredients to counter the inherent photochemical reaction, by decreasing free radical injuries. Antioxidants act by reversing the oxidative stress developed by UVR and infrared radiations. Their protective efficacies have been reported in various studies. Still larger studies are needed to confirm their level of protection and long term safety profile. Carotene, antimalarials, vitamin A, C and E, green tea extracts, selenium, retinol and many more have been reported to have photo protective qualities.[8] Three widely used and studied systemic sunscreens are polypodium leucotomos extract, afamelanotide and nicotinamide.

Polypodium leucotomos, belonging to fern group of plants, has been shown to decrease psoralen plus UVA and UVB phototoxicity.[16],[17] The oral administration of the drug as 240 mg twice a day provides a SPF value of 3-8, and has been shown it to be protective against erythemogenic spectra of UVR.[18]

The melanocortin-1 receptor agonist, afamelanotide exerts its photoprotection via increasing synthesis of eumelanin. It has been approved as an adjuvant in adult patients of erythropoietic protoporphyria by the European Union to prevent phototoxicity and has to be administered as 16 mg subcutaneously every 2 months.[19]

When taken orally, nicotinamide (active form of niacin) has been shown to be photo protective by enhancing intracellular adenosine triphosphate,[20] DNA repair and boosting cell energy.[21],[22] Its broad photoprotective effect against development of premalignant lesions is being studied currently.

Sunscreens related controversies:

1. Hormonal effect:

There are rising concerns about possible hormonal disruption of sunscreen agents benzophenones specially oxybenzone, which is in widespread use since 1970s. Initial in vitro studies have shown its antiandrogenic and estrogenic effects.[23],[24] However in recent in-vivo studies the claims were not substantiated about cause and effects.[25] Careful observation and further human studies are needed to clarify this aspect.

2. Role of antioxidants:

Nowadays, many sunscreen manufactures are using antioxidants beside active ingredient to decrease the adverse effects of free radicals, generated by UV exposure. In vivo studies have shown decreased matrix metalloproteinase-1 activity and less pigment induction with use of stabilized antioxidants[26] but a recent study has proven otherwise because of lack of stability of antioxidants used in sunscreens.[27]

3. Nanoparticulate sunscreen:

There are concerns about rising use of nanotechnology in sunscreens (to make it cosmetically elegant) as nanoparticles can produce free radicals on UV irradiation.[28] Various studies have shown that the confinement of nanoparticles is limited to stratum corneum. In addition, the use of coated nanoparticles has made it safe for usage in humans.[29] However application at sites with severely impaired barrier function should be minimized till further data are available.

4. Photocarcinogenic potential of retinyl palmitate (RP):

It is a storage form of vitamin A which was approved by the FDA to use in cosmetics and edibles. In view of rising concerns about photo carcinogenicity of compound, Wang et al[30] concluded in-depth review on this topic in addition to a large in-vivo study conducted by the FDA. None of the above could establish conclusive evidence about photo-carcinogenicity of RP. Also there is long standing history of safety profile of the product in humans.[11]

5. Vitamin D deficiency:

There have been concerns of vitamin D deficiency regarding universal use of sunscreens as ninety percent of vitamin D production in skin happens as a result of UV exposure. However, review of literature by Norval et al[31] concluded that deficiency doesn't occur with normal usage of sunscreen most likely due to insufficient application of sunscreen by most individuals. Unprotected exposure to UVR is not recommended to obtain vitamin D and supplementation in individuals at risk is advised as supported by the Institute of Medicine.[19]

6. Pediatric population and sunscreen:

Although there are no deleterious effects of sunscreen documented with use in early age, still it is advisable to use sunscreens containing inorganic filters over exposed area, in

adjunction to other sun protective measures.[11]

7. Environmental issues:

The effects of organic sunscreens on environment have become a burning issue since water sources are found to be contaminated with sunscreens specially oxybenzone in various studies.[32] They can react to chlorine in pools to form brominated transformation products which are hard to remove by usual water filters. In addition to this, studies have indicated possible role of oxybenzone in coral bleaching.[33] The adverse impact is still being studied.

8. Role of higher SPF:

The labeling restriction of SPF value more than 50 as SPF50+ by the US FDA has created a stir about significance of higher SPF. Various studies have shown that sunscreen with higher SPF has provided better protection against sunburn and UV induced phototoxicity.[34] The higher SPF can compensate the efficacy of a product in actual use as there are enough evidences to suggest that on an average only one third amount of a given SPF is attained due to insufficient and improper application.[14]

9. Sunscreen and special populations

There has always been confusion about prescription of sunscreen in Fitzpatrick skin type IV to VI as these skin types are less prone to sun damage because of inherent protective quality of melanin. However, enough evidence of photodamage including photo ageing has been documented.[35] In addition, malignant melanoma carries a poorer prognosis in POC (people of colour) despite low prevalence. Hence it is recommended to use regular sunscreen with other sun protective measures the same way as in other skin types. Broad spectrum sunscreen with SPF ≥30, specially containing inorganic filters are better suited for POC as they are more acceptable.[36]

Use of regular sunscreen with other photo protective behavior measures should be followed in patients of organ transplantation and dialysis to decrease the risk of premalignant and malignant changes in skin.[37]

10. Sunscreen and cosmetics:

Besides conventional sunscreen cream and lotion, now a day active ingredients are seen as foam, gel, mousse, spray, pastes, oils, butters, sticks and ointments. The pharmaceuticals and cosmetic giants are using sunscreen in range of over the counter products and cosmetics like foundation, compact, shampoo, lipstick, lip balm and wipes, with claim of varied SPF. The efficacy of these are not well established and not approved by the FDA yet. The spray forms are being promoted as convenient to use in children and over relatively non accessible sites like back in adults. For acne prone skin, gel and sprays forms are tolerated well. Sprays have also shown to retain active compounds to superficial layers of epidermis thus decreasing the risk of deeper penetration.[38]

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TRICHOSCOPIC FINDINGS IN VARIOUS SCALP ALOPECIAS

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Abstract

Background: Many patients of alopecia are encountered in our daily practice with diagnostic dilemma. Dermatoscopy is evolving as an important tool which aids our clinical diagnosis. **Aims:** To explore the utility of dermoscopy and to study the dermatoscopic findings in Cicatricial and Non Cicatricial Alopecias of the scalp. **Method:** An Observational study was conducted at rural based tertiary centre in Gujarat, India, during the period of June 2013 to May 2014 after getting ethical clearance. **Results:** Amongst 194 enrolled,142(73.12%) were males and 52(26.80%) were females. 45.36% were diagnosed to have Alopecia Areata(AA) with the most common finding being exclamation marks found in 75% patients.35.05% were diagnosed to have alopecia Androgenic (AGA) with the most common finding being thin and vellus hairs found in 83.82% patients.6.19% were diagnosed to have Psoriasis with red dots ,globules and silvery scales seen in 100% patients. 5.15% were diagnosed to have Tinea Capitis with corkscrew hairs seen in 100% patients. 3.6% were diagnosed to have Discoid Lupus Erythematosis with scattered dark brown discoloration of skin being the commonest finding was seen in 85.71 patients. 2.58% were diagnosed to have Sebborheic dermatisis with yellow scales seen in 100% patients. 2.06% were diagnosed to have Lichen Plano Pilaris with perifollicular inflammation seen in 100% patients. **Conclusions:** Dermoscopy has been shown to enable the visualization of sub-macroscopic morphologic structures invisible to the naked eye. Trichoscopy is used as a diagnostic aid in differential diagnosis of hair loss and scalp diseases.

Key Words- Trichoscopy, Alopecia, Lichen plano pilaris, Scalp Psoriasis, Seborrheic Dermatitis, Discoid Lupus erythematoses, Tinea Capitis

Introduction

A dermoscope (dermatoscope) is a non-invasive, diagnostic tool which visualizes subtle clinical patterns of skin lesions and subsurface skin structures not normally visible to the unaided eye. The basic principle of dermoscopy is transillumination of a lesion and studying it with a high magnification to visualize subtle features. [1] Some dermoscopic patterns are observed consistently with certain diseases and these then could be used for their diagnosis. Skin surface microscopy of the scalp is termed as Trichoscopy.

Structures which may be visualized by trichoscopy includeHair shafts, Hair follicle openings, Perifollicular epidermis, Cutaneous microvessels. Trichoscopy allows analyzing acquired and congenital hair shaft abnormalities[2,3,4]. Normal hair shafts are uniform in shape and color with continuous, interrupted, fragmented or absent medulla[5].About 10% of normal human scalp hairs are short, hypopigmented vellus hairs[3,4]. Trichoscopy may distinguish whether hair follicle openings are normal, empty, fibrotic or containing biological material, such as hyperkeratotic plugs or hair residues.

"Dots" is a common term for hair follicle openings seen by trichoscopy.[5]

Black dots ("cadaverized hairs") represents pigmented hairs broken or destroyed at scalp level.[5] They are observed in alopecia areata,[6] dissecting cellulitis, tinea capitis, chemotherapy - induced alopecia, and trichotillomania, but may be incidentally observed also in other diseases and after laser depilation or trichogram.

Yellow dots are follicular infundibula with keratotic material and/or sebum variable in color, shape and size.Yellow dots are present in alopecia areata(AA), [6] discoid lupus erythematosus (DLE) and androgenic alopecia(AGA). [7] The predominance of yellow dots in the frontal area compared to the occipital area favors the diagnosis of (female) AGA.[7]

White dots represent areas of perifollicular fibrosis and are observed most commonly in LPP. Another type of white dots, the small, regular pinpoint white dots are observed in sun exposed areas and in dark skin phototypes regardless of hair loss. They correspond to empty hair follicles or to the eccrine sweat duct openings.

Red dots represents blood vessels and is commonly seen in Psoriasis in "honeycomb pattern". Also seen in DLE and are believed to be a positive prognostic factor.[8]

Brown or brown-gray dot are the characteristic finding in the eyebrow area of patients with frontal fibrosing alopecia(FFA). It is a favorable prognostic factor for eyebrow regrowth.

The word "alopecia" comes from the Greek word "alopex" for "fox.", which means hair loss when afflicted with a skin disease (the "mange").Alopecia is defined as complete or partial loss of hair from scalp and other hair bearing sites of the body[9].

Male and female pattern baldness are androgen dependent alopecia. It is very common in males that about 80 %. In males, it clinically presents as recession of frontal harline, bitemporal baldness or a vertex pattern of hair loss. Occipital scalp is relatively spared. In females, pattern baldness is noticed more in the pubertal, perimenopausal and postmenopausal age groups; in the form of thinning of hair leading to widening of the central partition in a Christmas tree pattern. The diagnosis is mostly made clinically. In doubtful cases dermoscopy can help. The dermoscopic findings of AGA as variation of hair diameter more than 20% in all the cases and in late cases there can be a predominance of yellow dot[10] There is perifollicular pigmentation called peripilar sign in all patients but it could be documented only in 66 % of Asian patients probably because of the confounding skin colour[11].

Alopecia Areata commonly presents as round or oval patches of non-scarring hair loss. Short 'exclamation mark' hairs (i.e. distal end broader than the proximal end) can often be seen, particularly at the margins of areas of alopecia. Dermoscopy can be particularly useful to distinguish AA from other types of alopecia. The dermoscopic findings include yellow dots,black dots, broken hairs , short vellus hairs, tapering hairs, coudable and cadaverised hairs. [12]

Tinea Capitis is an infection of the scalp and hair caused by the dermatophytes Microsporum and Trichophyton. It is most common in infants and rare after puberty. Its dermatoscopic finding is characterized by a pigmented, homogeneously thickened and sharp slating ended hair shaft. The comma hairs and the corkscrew hairs appear to be specific dermatoscopic findings of dermatophytosis of the scalp[13], regardless of the etiological agent.

Seborrheic Dermatitis is a common type of non-atopic dermatitis with red, mildly itchy skin seen particularly on the scalp, T-zone of the face (especially the melo-labial fold) and skin folds, such as the axillae and groin. On Dermascopy seborrhoeic dermatitis (SD) was characterized by arborizing vessels and atypical red vessels[14]. Featureless areas devoid of any particular vascular patterns were also frequently observed. Scales were observed commonly, there was no significant difference in the frequency and characteristics of the scales when they were observed using dermoscopy.

Psoriasis is common erythematous-squamous dermatoses that may present with scaly erythematous patches on the scalp.The most significant dermoscopic features of scalp psoriasis were red dots and globules, twisted red loops, and glomerular vessels all arranged in honeycomb pattern[14].

DLE produces alopecia probably because of the inflammation of the infundibular region of the hair follicle. Patches on scalp are often itchy in the form of areas of scarring erythema and scaling with follicular plugging. In extensive cases: extensive areas may be involved resembling pseudopelade of Brocq. Involvement of the scalp commonly produces a scarring alopecia, [15] but there has been an increase in incidence of alopecia areata among patients with LE.[16] Dermoscopic findings described in DLE include peripilar erythema and scaling, White patch, branching capillaries, keratin plugs and decrease in number of follicular ostia.

Lichen planopilaris presents with patches of hair loss showing violaceous papules erythema and scaling of the underlying scalp.

Table 1: Types of alopecia with their causes

Cicatricial alopecia				Noncicatricial alopecia			
Disorder	Males	Females	total	Disorder	Males	Females	total
LPP	1	3	4(2.1%)	ALOPECIA AREATA	55	33	88(45.3%)
DLE	2	5	7(7%)	ANDROGENIC ALOPECIA	67	1	68(35.0%)
				PSORIASIS	8	4	12(6.2%)
				SEBORRHEIC DERMATITIS	4	1	5(2.6%)
				T. CAPITIS	5	5	10(5.2%)
Total: males-3, Females-8, Total- 11				Total	139	44	183

Papules get replaced later by follicular plugs and scarring. Hair pull test is positive in the margins.In the fibrotic stage of lichen planopilaris the dominating features are big, irregular (classic) white dots, which merge into milky-red (strawberry icecream color) or white areas.[17]

Aim

This study was conducted to study the dermatoscopic findings of various Cicatricial and Non Cicatricial scalp alopecias, To explore the utility of dermoscopy in the examination and diagnosis of various hair loss disorders .

Methods

The study was conducted at Skin opd, Shree Krishna Hospital during the period of June 2013 to May 2014 after getting ethical clearance from Institutional Human Resource and ethics Committee.

All consenting male and female patients, with alopecia, attending the Skin Department were enrolled. Hair loss occurring due to any external injury, chemotherapy, other drugs, any Systemic cause or Hair Shaft deformity were excluded.

194 patients were enrolled during the period of one year after written informed consent. In every case detailed history was elicited and clinical examination was carried out following which dermatoscopy was done with a non polarised dermatoscope to correlate our clinical findings and aid to our diagnosis. Diagnosis was done clinically and histopathological examination was performed whenever in doubt

Results

Total of 194 patiets of alopecia were enrolled out of which 142 (73.12%) were males and 52 (26.80%) were females.(table 1)The most common age group affected was 21-30 with 78(40.20%) patients. Total 88 (45.36%)patients were diagnosed to have AA.(Table 1) Most common dermatoscopic finding was exclamation marks seen in 66 (75%) patients followed by black dots seen in 65 (73.86%) patients. Caudability sign, sparing of white hairs can also be observed dermatoscopically. (Figures 1a,1b,1c) (Table2)

Total 68 patients (35.05%) were diagnosed to have AGA.(Table 1) Most common dermatoscopic finding was thin and vellus hairs seen in 57 (83.82%) patients, followed by yellow dots seen in 51 (75%) patients.(Figures 2a,2b) (Table 2) Family history was positive in 43 (63.24%) patients out of whom 36 (83.72%) patients had positive paternal history, 5 (11.63%) patients had

Table 2: Trichoscopic findings of various scalp alopecias

Disorder	Findings	Number(%)	Males(%)	Females(%)
Alopecia areata (N=88)	EXCLAMATION MARKS	66 (75)	39(70.91)	27(81.82)
	TAPERED HAIRS	60 (68.18)	41(74.55)	19(57.58)
	BLACK DOTS	65 (73.86)	42(76.36)	23(69.67)
	YELLOW DOTS	58 (65.90)	36(65.45)	22(66.67)
	UPRIGHT REGROWING HAIR	38 (43.18)	14(25.45)	24(72.73)
	VELLUS HAIRS	26 (29.54)	17(30.91)	9(27.27)
	BROKEN HAIRS	46 (52.27)	27(49.1)	19(57.58)
Androgenic alopecia (N=68)	THIN AND VELLUS HAIRS	57(83.82)	56(83.58)	1(100) (Female pattern)
	HAIR SHAFT THICKNESS HETEROGENEITY	47(69.11)	47(70.15)	0
	PERIFOLLICULAR DISCOLORATION	32(47.05)	32(46.27)	0
	YELLOW DOTS	51(75)	50(74.63)	1(100) (Female pattern)
Lichen plano pilaris (N=4)	SILVER WHITE PERIFOLLICULAR SCALING	3(75)	0	3(100)
	PERIFOLLICULAR INFLAMMATION	4(100)	1(100)	3(100)
	CONCENTRIC BLOOD VESSELS	1(25)	1(100)	0
	VIOLACEOUS BLUE INTERFOLLICULAR AREAS	1(25)	0	1(33.33)
	WHITE DOTS OVER RED BASE	3(75)	1(100)	2(66.67)
Discoid lupus erythematosis (N=7)	LARGE YELLOW DOTS	5(71.42)	1(50)	4(80)
	THICK ARBORIZING VESSELS	3(42.85)	1(50)	2(40)
	SCATTERED DARK BROWN DISCOLORATION OF SKIN	6(85.71)	2(100)	4(80)
Psoriasis (N=12)	RED DOTS AND GLOBULES	12(100)	8(100)	4(100)
	TWISTED RED LOOPS	7(58.33)	5(62.5)	2(50)
	SILVERY SCALES	12(100)	8(100)	4(100)
Sebborheic dermatitis (N=5)	THIN ARBORIZING VESSELS	3(60)	3(75)	0
	YELLOW SCALES	5(100)	4(100)	1(100)
Tinea capitis (N=10)	COMMA HAIRS	8(80)	3(60)	5(100)
	CORKSCREW	10(100)	5(100)	5(100)

positive maternal history, 2 (4.65%) patients had positive family history both paternally and maternally.

Amongst 194 enrolled, 4 (2.06%) were diagnosed to have LPP.(Table 1) Most common dermatoscopic finding was perifollicular inflammation seen in 4 (100%) patients, followed by silver white perifollicular scaling seen in 3 (75%) patients. (Figures 3) (Table 2)

Total 7 (3.6%) were diagnosed to have DLE.(Table 1) Most common dermatoscopic finding was scattered dark brown discoloration of skin seen in 6 (85.71) patients followed by large yellow dots seen in 5 (71.42) patients. (Figures 4a,4b) (Table2)

Out of 194 enrolled, 12 (6.19%) were diagnosed to have Psoriasis.(Table 1) Most common dermatoscopic finding was red dots and globules seen in 12 (100%) patients and silvery scales in 12 (100%) patients. (Figures 5a,5b) (Table 2)

Five patients were diagnosed to have SD.(Table1) Most common dermatoscopic finding was yellow scales seen in 5 (100%) patients followed by thin arborizing vessels seen in 3 (60%) patients. (Table2)

Total 10 (5.15%) out of 194 were diagnosed to have TC.(Table 1) Most common dermatoscopic finding was corkscrew hairs seen in 10 (100%) patients, followed by black dots seen in 9 (90%) patients. (table2).

Discussion:

Scalp alopecias reflect a broad spectrum of heterogeneous diseases and are among the most common dermatologic disorders. A careful history and a thorough clinical examination are usually adequate to establish the correct diagnosis. In some cases, eg, cicatricial alopecia, a scalp biopsy may be necessary. However, the histopathologic features are not always diagnostic[5]. Consequently, new diagnostic methods are required. Scalp dermatoscopy is a promising way to facilitate the diagnosis of scalp and hair disorders. [18] Trichoscopy may represent an important link between clinical and histologic diagnoses.[19]

Hair loss can have significant effects on patients' quality of life, and a prompt diagnosis of the different types of alopecias and an early intervention is needed. This review highlights the main dermoscopic findings in the different types of Scalp Dermatoses associated with alopecia.We believe that this important tool has demonstrated to help dermatologists in highlighting minute details thus avoiding unnecessary biopsies.

The prevalence of alopecia in our country has been measured for the major groups of Alopecias. But there is no literature regarding the percentage distribution of each diagnosis. AGA is considered to be the commonest cause of alopecia worldwide[20]. The approximate prevalence of AA worldwide is around 0.1-0.2%.[21] In our study the prevalence of alopecia was 0.58% of the out patient population. These two are considered as the commonest causes of alopecia. In a large hospital based study by Sharma et al in north India in 808 patients, AA patients comprised 0.7% of the outpatient population.[22] In our study, AA was the commonest diagnosis madewith 88 (0.26%) patients followed by AGA 68(0.20%) patients.

In our study, amongst 194 enrolled, 142 (73.12%) were males and 52 (26.80%) were females.The study conducted by Vivek V.et al at Sir Takhtasinh Hospital, Bhavnagar, India included a

Figure 1(a,b,c): **1a.** Dermatoscopy (50x) of alopecia areata showing Exclamation Mark Hair. **1b.** Dermatoscopy (200x) of alopecia areata showing sparing of White Hair. **1c.** Dermatoscopy (200x) of alopecia areata showing Coudability Sign.

Figure 2 a. Dermatoscopy (50x) of androgenic alopecia showing Thin and Vellus hair, Hair shaft thickness Heterogeneity

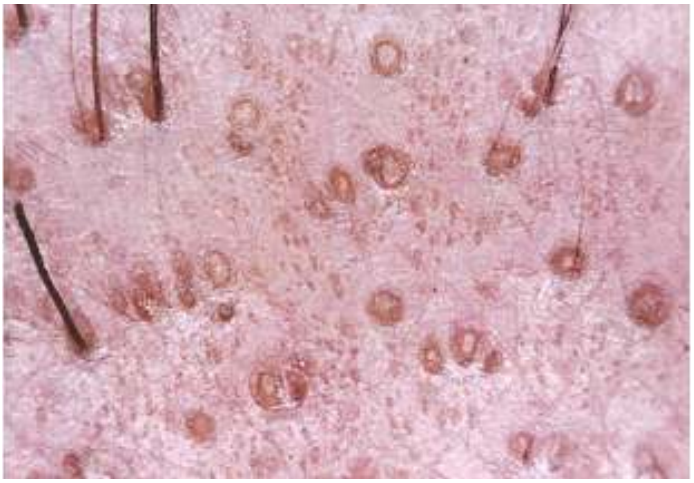

Figure 2 b. : Dermatoscopy (50x)of androgenic alopecia showing Empty hair follicles.

total 112 patients in 2 years out of which 58 patients (51.7%) were males and 54 patients (48.21%) were females. [23]

Most common dermatoscopic finding seen in our study was exclamation marks seen in 66 (75%) patients followed by black dots seen in 65 (73.86%) patients. The dermatoscopic finding in AA in our study shows sparing of gray hair which consolidates the established autoimmune theory of AA which states CD8 T cells are directed against follicular melanocytes.

Lacarrubba and colleagues[24] investigated 200 patients with alopecia areata, subdivided into acute and chronic disease. This study identified 3 features of acute AA: micro-exclamation marks, black dots, and vellus hairs. Inui and colleagues[6] identified similar markers of disease activity (black dots, tapering hairs, and broken hairs) based on trichoscopy performed in 300 patients with AA. In this study, vellus hairs were found to be a marker of longlasting, inactive disease. Ross and colleagues[18]divided 58 patients with AA into the following subgroups: patchy, ophiasis, diffuse, and alopecia totalis / universalis. Trichoscopy features were similarly expressed in all investigated subgroups. The authors' experience[9,25] shows that black dots are a most constant marker of disease activity in AA.

In our study males were commonly affected with male: female ratio of 1.66:1; in comparison with study conducted by Inui et al., where male: female ratio was 2.57:1.[6]

Exclamation marks were seen in 31.7% (95/300) cases of AA by Inui et al. [6] and 12.1% (8/66) cases by Mane et al. [26]Exclamation mark sign, which is specific finding of alopecia areata, was seen in 75% of patients in our study.

Black dots seen in our study was 65 (73.86%) patients while, Inui et al., [6]demonstrated Black dots in 44.3% (133/300) cases of AA and Mane et al. demonstrated Black dots in 67.7% (44/66). [26]Yellow dots were seen in 58 (65.90%) of cases in contrast to Ross et al.'s study where 94.8% cases with Alopecia had Yellow Dots(55/58 cases)[5]. Mane et al.,[26] reported an incidence of 81.8% among 66 patients.This differences could be explained by the shampooing practices of the patients.

Broken hairs were seen in 46 (52.27%) patients in our study, Inui et al. [6] demonstrated Dystrophic hairs in 45.7% (137/300) of alopecia cases. 55.4% patients had dystrophic hairs in the study conducted by Mane et al. [26] Our study findings are consistent with the above study findings.

Vellus hairs seen in 26 (29.54%) patients while study by Inui et al., [6] Small villous hairs (SVH) were observed in 72.7% (218/300) of cases. Mane et al., [26] demonstrated SVHs in 40.9% of patients.

AGA is characterized by hair diameter diversity due to miniaturization of the hair follicles. Variability in hair shafts diameter of more than 20% is diagnostic of this condition[25]. In early AGA, it is common to see peripilar brown depressions described as peripilar signs[27]. In patients with advanced androgenetic alopecia, yellow dots can be observed and the sun-exposed scalp often shows the honeycomb pigment pattern. Yellow Dots in AGA are thought to be the result of sebaceous hypertrophy and lagooning in glands as a result of end-organ hypersensitivity[7]. The characteristic trichoscopic findings of AGA are known as Heterogenous hair density and Perfollicular pigmentation[4] Perifolliculr pigmentation is thought to be the result of dermal infiltrates in AGA[25,27]

Total 68 (35.05%) were diagnosed to have AGA. Hair shaft thickness heterogeneity, tapered hairs were seen in 47(69.11%)patients, while study by Inui et al, Hair Shaft Thickness heterogenicity and thin hair was observed in 100% of patients and Yellow Dots were seen in 25.4% of patients. Ross et al [5]emphasized Yellow Dots being higher in late AGA in their

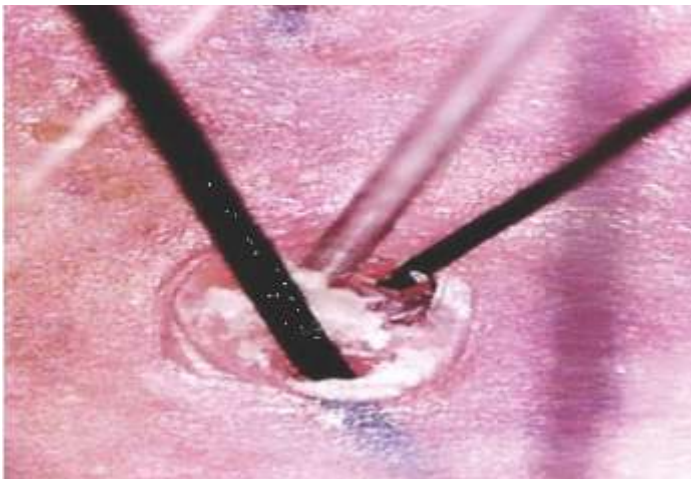

Figure 3. : Dermatoscopy (200x) of LPP showing Silver White Perifollicular Scaling

Figure 4 a. (50x) of DLE showing Thick Scaling with Yellow Dots

Figure 4 b. : Dermatoscopy (200x) of DLE showing Thick Arborizing Vessels (with paraffin)

study and Lacarrubba et al.[24] underlined miniaturization as being higher in early AGA.

Perifolliculr pigmentation was first described by Deloche et al.[27]Inui et al.[28]reported Perifollicular pigmentation in almost all patients with alopecia who had fair skin. While in our study perifollicular pigmentation was seen in only 2(47.05%) patients as the study was done in Asian population.

In our study Family history was positive in 43 (63.24%) patients. Sawant et al reported that 51.3 % of the patients of androgenic alopecia had Positive family History at the time of presentation.[29]

LPP commonly affects women and its variant FFA significantly affects older women. The typical age of onset is 40 - 60 years.In LPP, dermoscopy reveals absence of follicular openings and the presence of characteristic perifollicular scales (peripilar casts) at the periphery of the patch. Perifollicular erythema characterized by the presence of arborizing vessels around the follicular ostia is also observed. Blue-grey dots may be found in some patients, especially those with dark skin. A peculiar pattern of round perifollicular blue-grey dots "target pattern" may be observed in some dark patients with LPP.[30] Usually, LPP spares some terminal hair follicles inside the alopecic patches.[31]

Most common dermatoscopic finding in patients of LPP was perifollicular inflammation seen in 4 (100%) patients followed by silver white perifollicular scaling seen in 3 (75%) patients. As per a study Duque Estrada et al, Perifollicular scales were seen in 100% of patients with no patient having perifollicular inflammation. Coiled capillaries and branching capillaries were not seen in any patient. Pigment network and white patches were seen in 50% patients and Blue gray dots were seen in 25% of patients.[32]

DLE is considered as a chronic form of cutaneous LE and may be seen alone or as a part of SLE. It is the commonest form of cutaneous LE.

The most characteristic trichoscopy features of DLE of the scalp are thick arborizing vessels and large yellow dots. Scattered brown discoloration of the skin may be observed in some patients. Yellow dots with radial, thin arborizing vessels emerging from the dot are considered characteristic for discoid lupus erythematosus. This feature is sometimes referred to as "red spider in yellow dot."[9]Red dots, described by Tosti and colleagues,[8] are considered a good prognostic factor for of hair regrowth.

Most common dermatoscopic finding in patients of DLE was scattered dark brown discoloration of skin seen in 6 (85.71) patients followed by large yellow dots seen in 5 (71.42%) patients.

In Duque-Estrada et al study, Perifollicular scales and inflammation (0%) were not seen in any cases of DLE while Coiled

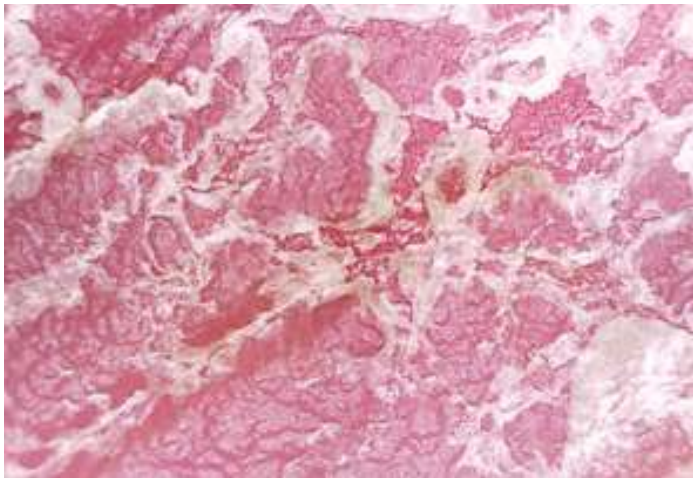

Figure 5 a. : Dermatoscopy (200x) of psoriasis showing Silvery Scales.

Figure 5 b. : Dermatoscopy (50x) of psoriasis showing Red Dots and Globules in Honeycomb Pattern after removing scales (with paraffin)

capillaries and Branching capillaries were seen in 20% and 100% cases. Pigment Network and blue gray dots were observed in 40% of patients. [30]our findings are consistent with the above study.

The most significant findings scalp psoriasis are: red dots and globules, twisted red loops, and glomerular vessels. In scalp psoriasis, within the typical scaly plaques, twisted red loops are observed at higher magnification. Twisted loops are also seen to a limited extent in unaffected psoriatic scalp as well as in newly treated psoriatic scalp.

Most common dermatoscopic finding was red dots and globules and silvery scales in all 12 patients of psoriasis followed by twisted red loops in 7 (58.33%) patients in our study.

According to Ross, Dermatoscopy is very useful to distinguish scalp seborrheic dermatitis form scalp psoriasis based on the vascular pattern. [5,33]. The vascular pattern of psoriasis is characterizedby twisted loops (100% of the cases) [5]. Kimet al[34]recently evaluated with dermatoscopy 55 patients with scalp psoriasis. Inpsoriasis they describe red dots and globules (mostcommon in their series), twisted red loops, and glomerular vessels. It is, however, important to keep in mind that twisted loops appear as dots on low magnification (up to 320). [35]

Seborrheic Dermatitis (SD)is characterized by presence of thin arborizing vessels and atypical red vessels. In seborrheic dermatitis, arborizing red lines, which have a wider caliber than the loops, can be observed. This may be helpful in differentiating it from psoriasis. However, capillary loop density seems to be similar in patients with psoriasis, SD and healthy scalp skin, and sometimes twisted loops are observed in sebopsoriasis like forms of SD[5]

.Most common dermatoscopic finding in SD was yellow scales seen in 5(100%) patients followed by thin arborizing vessels seen in 3 (60%) patients in our study.

Kimet al recently evaluated dermatoscopy of 41 patients withSD.In SD the most common patterns are arborizingvessels and atypical red vessels in the absence of red dots and globules. [34]

Tinea Capitis(TC) usually affects children and is commonest in the 3 -7 year age group. It rarely affects adults and has a female preponderance.Comma hairs and corkscrew hairs were found to be characteristic for TC. They have to be distinguished from corkscrew hairs observed in ectodermal dysplasias. Additional findings in TC are broken hairs, damaged hairs and black dots.

According to a study by Slowinska, In tinea capitis distinctive dermoscopic markers include black dots, broken hair and comma hair.

In our study most common dermatoscopic finding was corkscrew hairs seen in 10 (100%) patients followed by black dots seen in 9 (90 %) patients, comma hairs in 8 (80 %) patients, broken hairs in 7 (70 %) patients. Slowinska et al. reported comma hair in 80 % to be a distinctive marker for TC, followed by broken (66 %) and black dots (dystrophic hair) (60 %).[36] Comma hair, corkscrew hair and pigtail hair[37]were all observed only in patients of tinea capitis, thus forming specific features.

Conclusion: Dermoscopy has been shown to enable the visualization of sub-macroscopic morphologic structures invisible to the naked eye. Trichoscopy is used as a diagnostic aid in differential diagnosis of hair loss and scalp diseases. Hair and scalp disease show distinct patterns in most cases narrowing down the clinical differential diagnosis. Certain aspects of hair and scalp disorders can be better appreciated with dermoscopy than with the naked eye thus avoiding invasive procedures like Biopsy.

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LETTER TO EDITOR
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ARMOURED KELOID

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Sir,

Sir 1,2,3A 56 years old female presented with hyperpigmented to skin coloured leathery plaque on the chest covering whole of her chest region just like a armour and left scapular area from the last ten years [fig-1, 2]. Similar hypertrophic lesion was present on left ear lobule almost obliterating it [fig-3]. Patient complained of severe itching and pain over the lesions. Clinical diagnosis of keloid was made and patient was advised biopsy, which she refused. Patient was given intralesional steroid injections on prominent borders twice and later on she was lost to follow-up.

Keloids are abnormal tissue response to cutaneous injury. They are benign fibro-collagenous growths that rise above the skin surface and extend beyond the borders of the original wound. They have a tendency to occur in areas of wound healing with increased tension such as chest, deltoid and back. They may also rarely regress spontaneously and show a high level of recurrence after treatment[1] and are known to be notorious for their poor response to treatment owing to complex and ill-deciphered pathophysiology. Recent studies indicate that transforming growth factor beta and platelet-derived growth factor play an integral role in the formation of keloids.[2] Diagnosis is based on history and clinical examination and is confirmed by histopathology. Treatment modalities include but are not limited to simple excision, intralesional excision, local irradiation, steroid therapy, pressure therapy, cryotherapy, silicone gel application and enzyme therapy, alone or in combination. No single modality is 100% effective and recurrence rates range from 50% to 100%.3

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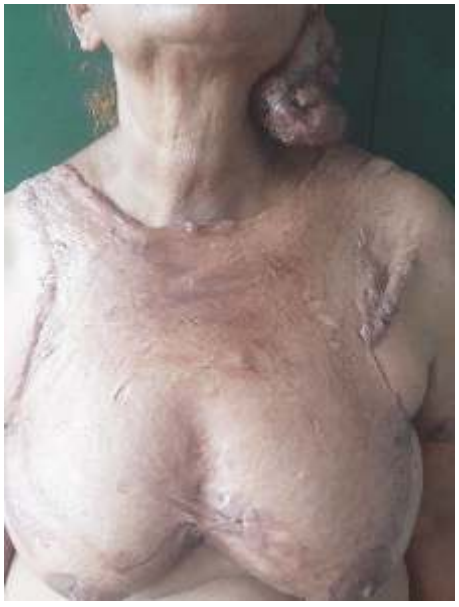

Figure 1. showing armoured keloid covering whole chest.

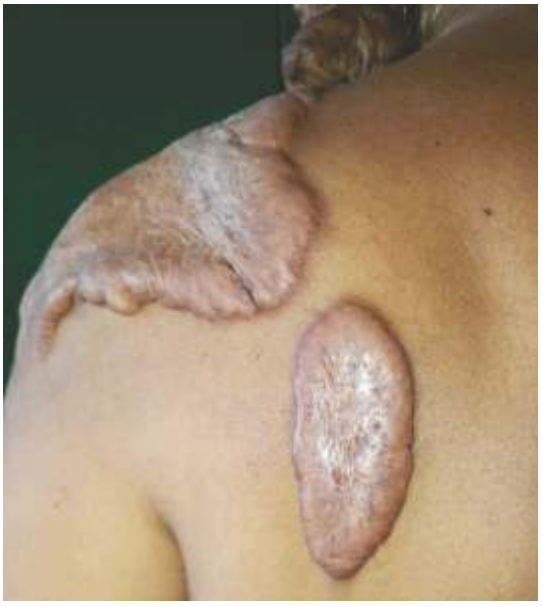

Figure 2. showing keloid on left scapular area.

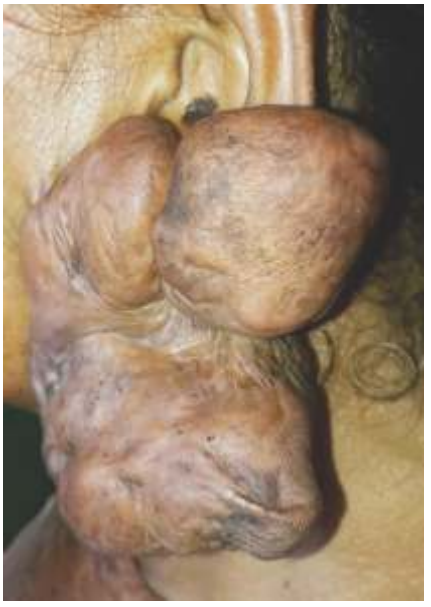

Figure 3. showing gaint keloid on left ear lobule almost obliterating it.

LETTER TO EDITOR
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UNILATERAL HYPERTROPHIC LICHEN PLANUS ON SOLE- A RARE CASE REPORT

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Sir,

Lichen planus (LP) is a common papulosquamous inflammatory dermatosis that can affect the skin, mucous membranes, hair and nails. Lichen planus is classically characterized by violaceous, scaly, flat-topped, polygonal papules to plaques and commonly involves the flexor aspects of the wrists, legs, oral and genital mucous membranes.[1,2]

Lichen planus has many morphological variants among which hypertrophic lichen planus is one of the type. It is an extremely pruritic form of LP and is characterized by presence of hyperkeratotic plaques which are usually seen over the shin and ankles. We are reporting a case of unilateral hypertrophic lichen planus in a patient who presented with mildly itchy hypertrophic plaque over right sole since past 6 years sparing other sites which is very unusual in presentation.

A 18 year old patient , student by profession presented with chief complaint of single raised lesion over right sole associated with mild itching since last 6 year. It started as a small asymptomatic pea sized lesion which gradually increased in size and developed mild itching over past 3 year. There is no history of pain or discharge from the lesion. There was no history of any palmer, scalp, oral or genital lesions.

There was no history of fever, weight loss, cough or joint pain. Personal or family history of atopy was absent. History of trauma, or any chronic illness were absent.

His general physical and systemic examination were normal. On cutaneous examination a single well defined erythematous plaque of size 5 X 4 cm with white firm scaling was present over the plantar aspect of right forefoot (Figure 1). Lesion was non tender and does not discharge or bleed on manipulation. Other body sites were spared. Oral mucosa , genital mucosa, nails and scalp examination were normal.

With this clinical feature we kept our differential as plantar psoriasis, hypertrophic lichen planus, tinea pedis, cutaneous tuberculosis and chromoblastomycosis.

Routine blood investigation including complete blood counts, renal function tests, liver function tests and thyroid profile were normal. Mantoux test was negative. Skin scrapping for KOH and fungal culture were negative. Tissue stain like PAS for fungus and AFB for TB were negative. X-ray chest did not revealed any abnormality.

Skin biopsy showed epidermal hyperplasia with foci of spongiosis and parakeratosis. There was moderately patchy perivascular & periappendigeal infiltrate of lymphocytes, plasma cells and histiocytes with occasional epitheloid cells .The reticular dermis showed an increased number of thick walled capillaries. Collagen bundles in papillary as well as reticular dermis showed thickening and haphazard arrangement. These findings were consistent with hypertrophic lichen planus.

On the basis of clinical feature and histopathological examination a diagnosis of hypertrophic LP was made and patient was started with oral and topical steroid. A significant improvement was seen within 3 weeks of treatment and dose of oral steroid was gradually tapered. Lesion healed completely in 3 months and patient is still under follow up without any reccurence for past 1 year.

The term 'LICHEN PLANUS' was coined by Erasmus Wilson in 1869.[3] Lichen planus is an chronic inflammatory dermatosis which produces a characterstic polygonal, violaceous pruritic papule and plaque with fine white reticulate streaks on its surface known as wickhams striae.[4] Incidence of LP varies from 0.1 % to 4 % depending upon the population studied.[5,6] In India different studies have reported that the incidence among dermatology outpatients is 0.38% to 1.4%.[7,8,9] It is commonly observed in patients of age group 31-40 years with minor

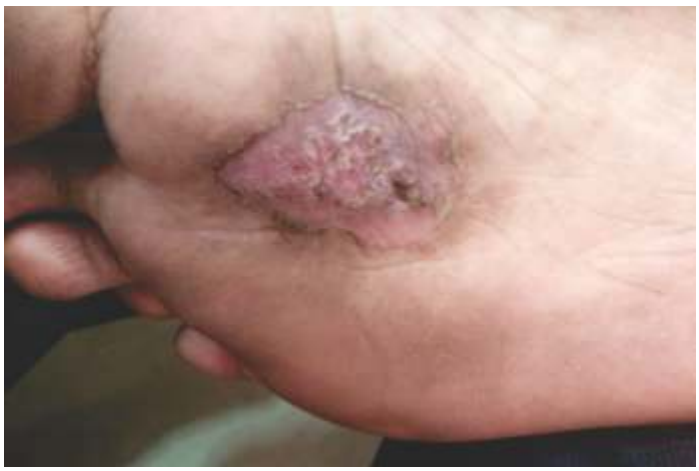

Figure 1: Erythematous plaque with white firm scaling over plantar aspect of right forefoot

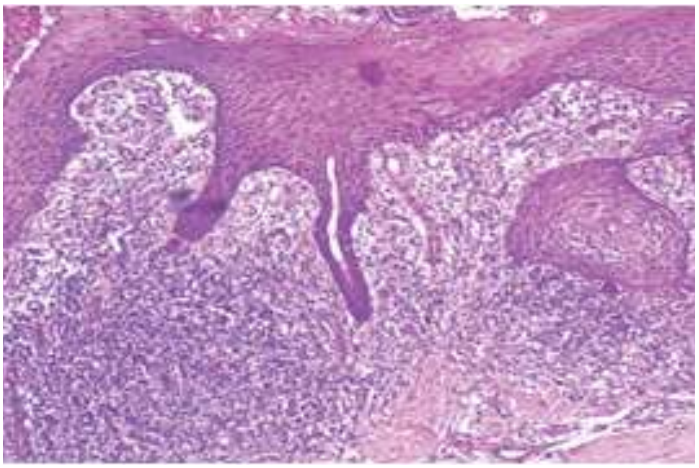

Figure 1: Epidermal hyperplasia with foci of spongiosis and parakeratosis

female predominance.[10,11]

Exact etiology of lichen planus is not known and multiple etiological factors have been associated with it. Genetic susceptibility plays a role especially in idiopathic LP. It is also supposed to be an immunological mediated disorder. Some cases of lichen planus are associated with autoimmune disease like myasthenia gravis , alopecia areata , lupus erythematosus whereas some are associated with infections like hepatitis B, hepatitis C and chronic active hepatitis.[12,13]

The lesions of classical LP involves flexural sites like arm, leg, trunk with sparing of face, scalp, palm & sole. Multiple morphological variants of lichen planus have been described like annular atrophic, bullous, erosive, hypertrophic, follicular etc.[14,15] Hypertrophic variant is severely itchy and commonly involves ankle and shin area.

Palmoplantar lichen planus is a rare localized variant of LP and usually lack the classical clinical morphology and becomes difficult to diagnosis sometime. In a study, palmoplantar LP together with accompanying skin involvement accounted for 26%.2 Classically palmoplantar LP present with pruritic well defined erythematous scaly or hyperkeratotic plaque followed by rare presentation of punctate keratoderma, diffuse keratoderma and ulcerated lesion.[16] In a study done by Sanchiz et-al the lesions of LP were more frequently present on soles than on the palm and common site of involvement is inner plantar arch. They also observed hyperkeratotic lesions in 25% patients.

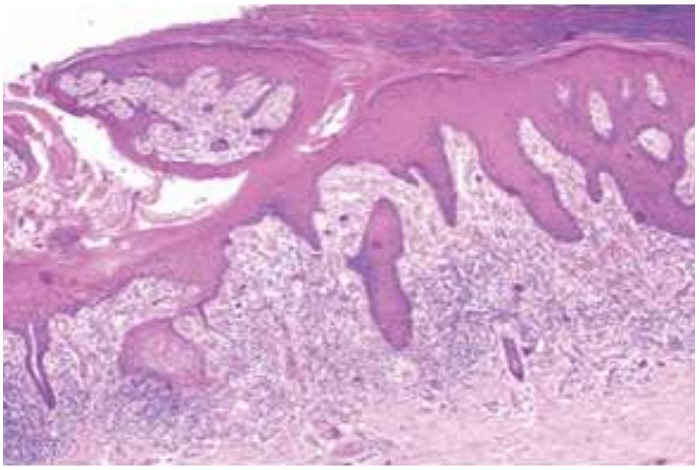

Figure 3: perivascular & periappendigeal infiltrate of lymphocytes, plasma cells and histiocytes with occasional epitheloid cells

In the current case the unilateral presentation of mildly itchy hyperkeratotic scaly lesion in planter area makes it very unique and interesting. As it lacks the classical presentation and wickham's striae, histopathology plays a significant role in confirming the diagnosis. Palmoplantar LP lesions usually heals spontaneously over a few months[17,18,19]. But in our case it lasted for 6 years without any improvement. Altman and Perry describe recurrence in 17% of patient, with an average duration of 8 months but in our case there was no recurrence during 1 year of follow up.[20]

The first line treatment of PPLP is topical or systemic corticosteroid. Other treatment modalities are topical tazarotene, oral cyclosporine, acitretin and phototherapy. The present case showed complete clearance with topical & oral steroids given for 3 months.

To conclude the PPLP is an uncommon variant and can present with clinical challenge to diagnose it early. We are reporting this case of unilateral hypertrophic lichen planus over sole because of its rarity.

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LETTER TO EDITOR
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UNILATERAL TRUNCAL ACNE AFTER LAMINECTOMY

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Sir,

Unilateral acne and related disorders (rosacea, seborrheic dermatitis, and demodicidosis) have been described in relation to paralyzed areas of different causes. We report a case of unilateral acne following laminectomy.

A 35-year-old male patient presented with papules and pustules predominantly located on one left half of trunk. The patient gave a history of an operation done in the neck region for cervical pain. He had undergone laminectomy for intradural extramedullarylesion in spinal canal at C7 level. Two weeks later, acne lesions appeared on left half of back. On examination there were pustular lesions present over back. The majority lesions were on the left half and a few on the other side. There were no lesions on face, chest and scalp. There was no history of any occlusive dressing in that area or any application of any medication. On further evaluation it was found that there was hypoaesthesia in the left side of back.

There have been several earlier reports of acne occurring in an unusual distribution.

'Immobility acne' occurring in the perioral region, following prolonged dental splinting after periodontal surgery[1]. Frictional acne in concert violinists[2] and in those wearing headbands.[3] The mechanism in these cases is possibly a hyperkeratinization response to local trauma or increased hydration of the pilosebaceous keratin. In cases of paralysis, including cases of Parkinson's disease and spinal cord injury, it has been suggested that an increased sebum excretion rate and the immobility of the affected area are most likely what caused the unilateral acne lesions. Seborrhoea is frequently seen in Parkinson's disease. This is associated with a raised Sebum Excretion Rate (SER) and it has been noted that following treatment with L-dopa, the SER was significantly reduced[3,4]. In a study done by Thomas et at, it was shown that the SER on the forehead is not significantly different in paraplegics and control subjects, but that the SER below the neurological lesion in paraplegic subjects is significantly greater than normal(P<o-oo1)[5]. Cases with unilateral acne have also been reported earlier post facial nerve palsy.[6,7,8] Thus in this case the cause for unilateral acne lesions might be due to an increased sebum excretion post laminectomy.

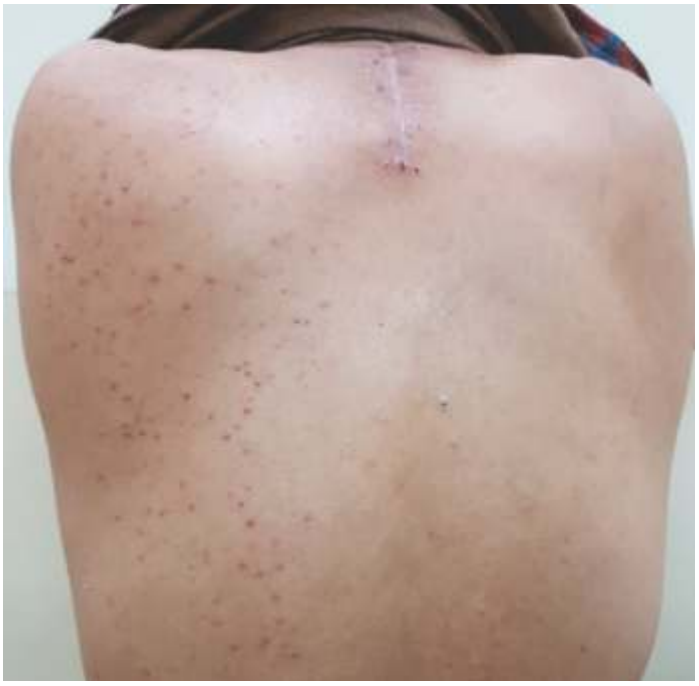

Figure 1: Unilateral truncal acne

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CASE REPORT
Year: 2019 • Volume: 2 • Issue: 1 • Page: 19-21

HAND, FOOT AND MOUTH DISEASE IN AN IMMUNOCOMPETENT ADULT- A RARITY

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Abstract

Hand, foot and mouth disease (HFMD) is a highly contagious viral infection in childhood or immunocompromised adults manifesting with low-grade fever along with vesicles or papules over oral mucosa, palms, dorsa of feet and buttocks. A 24-year old male presented with complains of fever since 3 days followed by lesions in mouth and over palms and soles for 2 days with history of malaise and throat pain. On examination there were multiple, discrete, ill-defined erythematous maculopapular rash present over palms, dorsa of feet and soft palate. Routine investigations were within normal limits along with seronegativity for HIV and syphilis. Symptomatic treatment resulted in resolving of lesions. HFMD is predominantly a childhood or immunodeficiency-associated disease spreading through feco-oral, oro-oral and fluids of the vesicles. Our case was a male in his mid twenties, suggesting that it can occur in immunocompetent adult also without any history of close contact.

Key words: Hand, foot and mouth disease, immunocompetent adult, viral infection

Introduction

Hand, foot and mouth disease (HFMD) is a highly contagious viral infection in childhood or immunocompromised adults. HFMD was clinically defined as any patient with an acute onset of vesicular eruptions on the hands and feet in association with oral sores.[1] Coxsackievirus A16 and enterovirus 71 being the most common cause of HFMD[2], CVA6 has also been identified in adult patients.[3-5] Here we report a case of an immunocompetent male with vesicular eruption and oral manifestations diagnosed as having HFMD

Case Report

A 24 year old male presented to skin opd in monsoon season with complains of fever since 3 days followed by lesions in mouth,palms and dorsa of feet for 2 days with history of malaise and throat pain. No history of herpes simplex infection, history of drug intake within the preceding 2 weeks (prior to the onset of skin lesions) were noted and no history of close contact with any infected child or adult was given. On examination there were multiple, discrete, ill-defined erythematous maculopapular lesions present over palms, dorsa of feet and soft palate. (Fig 1,2,3) Multiple clear fluid filled vesicles over bilateral ears (fig 4), multiple pustular lesions with crusting over buttocks. The results of hematological investigations were normal along with seronegativity for HIV and syphilis and Tzanck smears showed nonspecific inflammatory cells. Laboratory confirmationby electron microscopy of the pathogen causing the disease was not done due to resource constraints.Symptomatic treatment resulted in resolving of lesions.

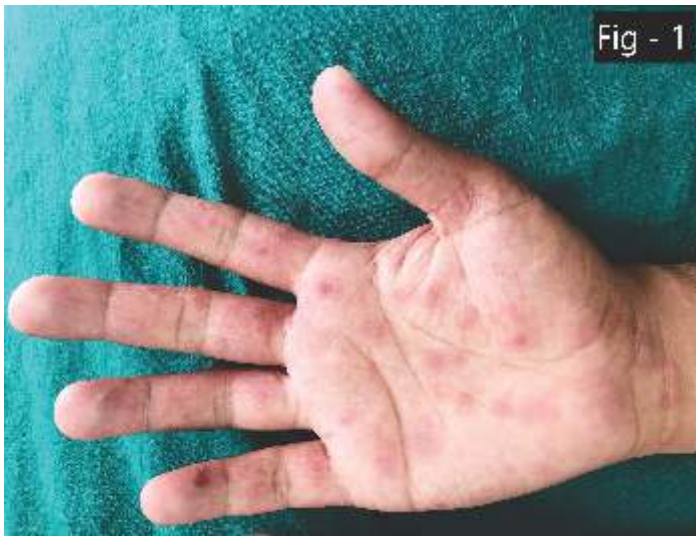

Figure 1 : multiple, discrete, ill-defined erythematous macule and papules present over palms.

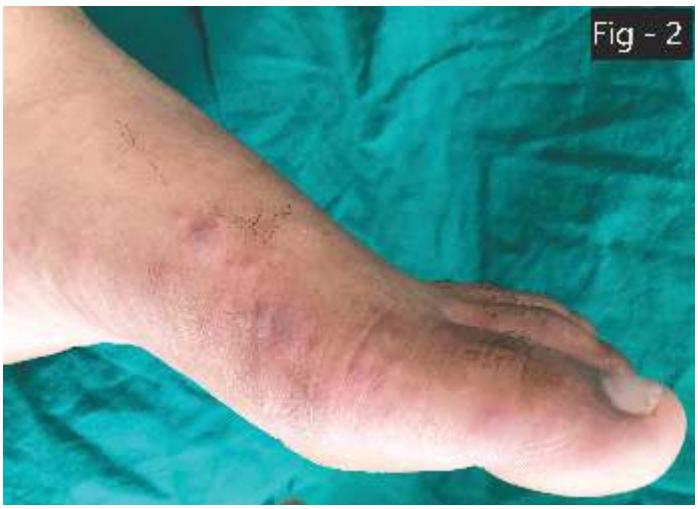

Figure 2 : multiple, discrete, ill-defined erythematous macule and papules present over dorsa of feet.

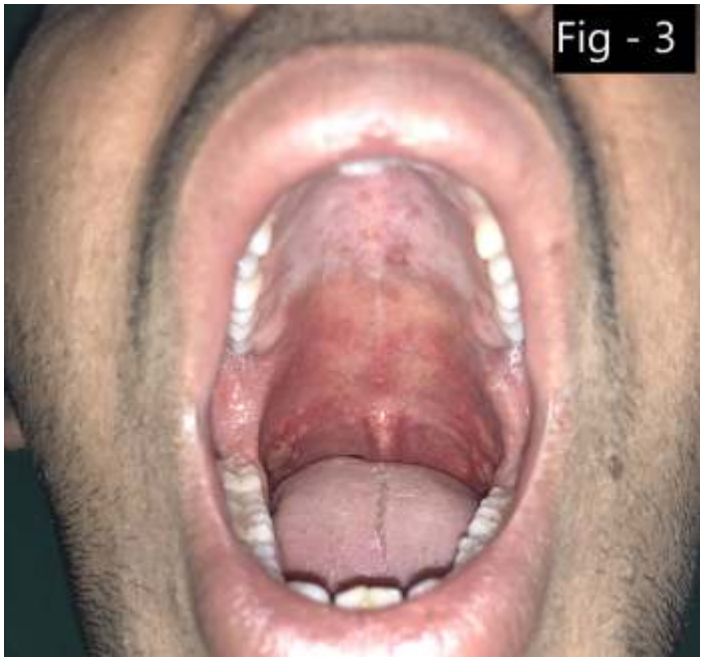

Figure 3 : multiple, discrete, ill-defined erythematous macule and papules present soft palate

Discussion

HFMD was clinically defined as any patient with an acute onset of vesicular eruptions on the hands and feet in association with oral sores.[1] HFMD predominantly occurs in children less than 10 years of age [6] and in immunocompromised adults, but can rarely be seen in immunocompetent adults. Coxsackie has been considered the most common cause of HFMD but traces of A5,A10 and not uncommonly human enterovirus 71 have been found.[2]Most cases are in young children during the autumn months, but outbreaks have occurred in communities of adults. Certain reports have shown that 25% adults were affected in several outbreaks in western countries between 2011 and 2012.[4] The disease is usually mild with an incubation period of 5-7 days, and lasting for about 7 days. In children it can be asymptomatic but in adults it can present with painful stomatitis. Fever accompanies the lesions. Skin lesions are mainly small vesicles upto 5mm in diameter, thin walled, pearly grey, most commonly over hands. Lesions heal spontaneously in 2 weeks without scarring. Atypical HFMD presents with more variable and severe manifestation such as diffuse rash, purpuric lesions and adult-age predilection.[7,8]A clinical diagnosis usually suffice the proof for HFMD but if histology is performed it usually shows spongiosis, intraepidermal splits progressing to vesicle formation, mononuclear cells entering the epidermis, and necrosis of individual keratinocytes. Laboratory tests include microneutralization test, reverse Transcriptase – Polymerase Chain Reaction, culture method, neutralizing antibody detection, and enzyme-linked immunosorbent assay.[9,10] Though HFMD is a self-limiting disease there should be surveillance maintenance as there is no effective chemoprophylaxis or vaccine available. Treatment is symptomatic. Maintenance of personal hygiene, social distancing and disinfection of the environment are probably the most effective measures to avoid spreading of the disease to the community. To our knowledge, only a few HFMD cases have been described in the literature in immunocompetent adults.[9,11,12]Public health personnel should be enabled proper epidemiological knowledge of HFMD to predict outbreaks of the disease and implement effective interventions so as to reduce the burden of disease in the society.

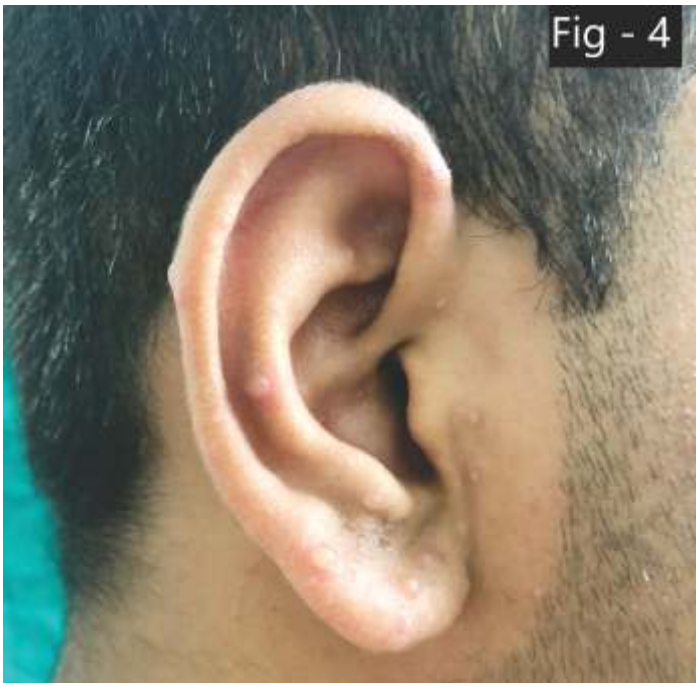

Figure 4 : Multiple clear fluid filled vesicles over bilateral ears

Conclusion

The current scenario states that the diagnosis of HFMD should be kept in mind whenever a patient presents with vesicles or maculopapular rash over palms, soles and oral mucosa to prevent its spread and complications, as it can rarely affect immunocompetent adults also.

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Modasia K, Shah A, Patel T, Tandel J, Vora R. Hand, foot and mouth disease in an immunocompetent adult- Ararity. JDA Indian Journal of Clinical Dermatology 2019;2:19-21.

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CASE REPORT
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A CASE OF DERMATOPATHIA PIGMENTOSA RETICULARIS

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Abstract

Dermatopathia pigmentosa reticularis is a rare ectodermal dysplasia with a triad of generalized reticulate hyperpigmentation, noncicatricial alopecia, and onychodystrophy. We report a case of a 3 year old female with reticulate hyperpigmentation present over whole body including oral mucosa and sclera. Diffuse thinning of hair on scalp was present. Poorly developed dermatoglyphics were there. There was onychodystrophy. Histopathology revealed superficial perivascular lymphocytic infilterate which also supported the diagnosis. There was no evidence of involvement of other ectodermally derived organ.

Key words: Ectodermal dysplasia, Reticulate pigmentation, Dermatoglyphics

Introduction

First described by Hauss and Oberste-Lehn [1] in 1958, Dermatopathia Pigmentosa Reticularis (DPR) is a rare ectodermal disorder. The diagnostic triad includes generalized reticulate hyperpigmentation, noncicatricial alopecia and onychodystrophy.

Case Report

A 3 year old female child, born out of non consanguineous marriage presented with darkening of skin since 6 months of age, which increased progressively to whole of the body including oral mucosa and sclera within a span of 1 month and diffuse thinning of hair on scalp. There was no history of photophobia. Hearing and sweating were normal. There was no history of similar illness in the family. She was born to a primigravida by normal vaginal delivery. She had one male sibling who was normal. The morphology of the hair shaft was normal on clinical and microscopic examination. Her developmental milestones were below 3rd centile. On examination, generalized reticulate hyperpigmentation was present (Figure No.1). Scalp hair was short and there was diffuse thinning. Nails were dystrophic. Poorly developed dermatoglyphics were there (Figure No.2).

Figure 1 : Diffuse reticulate hyperpigmentation of skin.

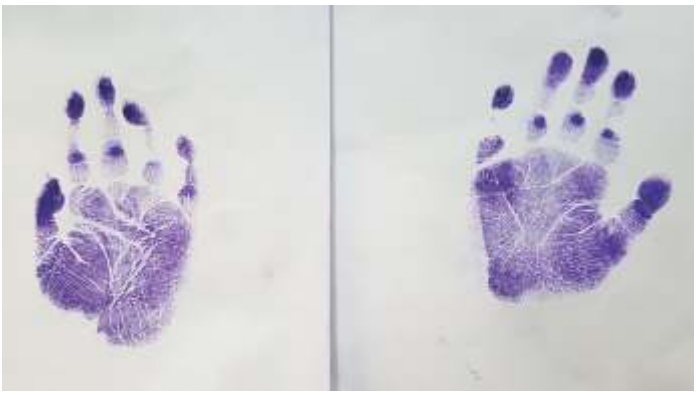

Figure 2 : Poorly developed dermatoglyphics.

Oral mucosa and sclera showed reticular pigmentation and teeth showed mineralization defect (Figure No.3). Her intelligence quotient was estimated to be in the normal range. Routine investigations in the form of complete hemogram, liver function test, renal function test and chest radiography were normal. Her thyroid profile was deranged and she was diagnosed with juvenile hypothyroidism. Her adrenal function was normal.

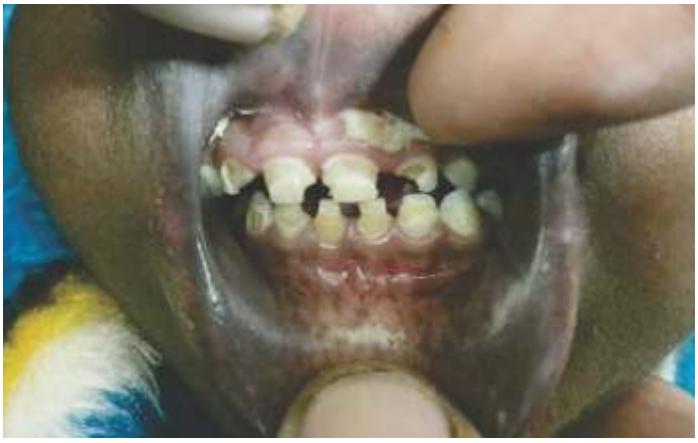

Figure 3 : Teeth mineralization defect.

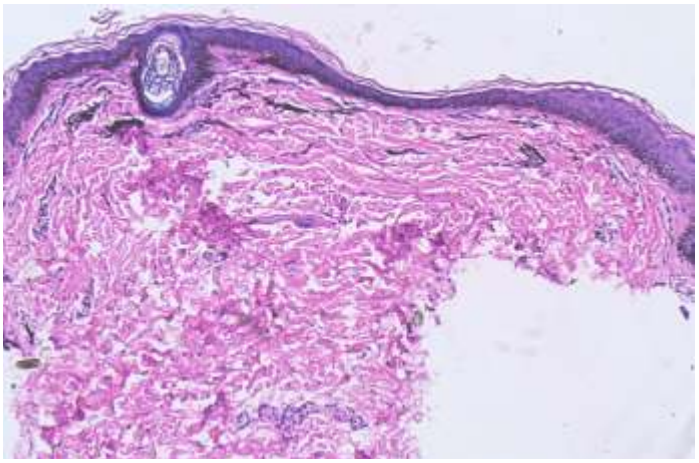

Figure 4 : superficial perivascular lymphocytic infilterate. The papillary dermis was slightly thickened with fibroblast and mucin.

Histopathology revealed superficial perivascular lymphocytic infilterate. The papillary dermis was slightly thickened with fibroblast and mucin. The epidermis was flattened at places (Figure No.4).

Discussion

DPR is an autosomal dominant ectodermal dysplasia. It usually presents as triad of reticular pigmentation, non-scarring alopecia and nail changes. The reticular pigmentation of DPR occurs at birth or during early childhood and persists throughout life.[2] Many other dermatologic findings have been associated with this triad, which include absent or decreased dermatoglyphia, hypohidrosis or hyperhidrosis, palmoplantar hyperkeratosis, acral nonscarring blisters, diffuse or punctate palmoplantar hyperkeratosis, darkly pigmented nipples, mucosal pigmentation, digital fibromatosis, neurofibromas, and wiry scalp hair.[3] Few extracutaneous manifestations that have been reported in the literature include fine punctate superficial spots in the cornea, Salzmann's nodular degeneration of the cornea, and early-onset gastric carcinoma.[4].

There are less than 20 cases of true DPR reported in the literature. Most of the cases were reported in Europe, and a few cases were reported in the USA and Asia; there was no race or sex predilection for DPR. The onset of the reticulate pigmentation of DPR usually occurs at birth or during early childhood, and the rest of its manifestations appear later. This condition should be differentiated from other genodermatosis associated with generalized reticulate pigmentation like dyskeratosis congenita (DKC), Naegeli-Franceschetti-Jadassohn Syndrome (NFJS), Dowling-Degos disease, reticulate acropigmentation of Kitamura and Haber's syndrome. Reticulate hyperpigmentation, mucosal leukoplakia, bone marrow dysfunction, cytogenetic instability, and a predisposition to malignancy are characteristic of DKC. These patients can have dental findings, reticulate hyperpigmentation, adermatoglyphia, palmoplantar hyperkeratosis, and nail anomalies similar to NFJS and DPR patients.[5] In Kitamura's disease, palmar pits and breakage in palmar ridges and acral hyperpigmentation, especially on the backs of hands and feet can be observed. Haber's syndrome is characterized by verruciform papular lesions of the trunk and a distinct facial erythema and telangiectasia, most commonly presenting in childhood. In X-linked reticulate pigmentary disorder in females, pigmentation occurs along the Blashcko's line.. Flexural reticulate hyperpigmentation occurs in Dowling-Degos disease. Additional findings, such as dark hyperkeratotic follicles, pitted perioral scars, and comedo-like lesions may occur. Galli-Galli disease also shows macular and papular reticulate pigmentation of flexures.The histopathology of the reticulate pigmentation of DPR is not diagnostic, and the reported histopathological features include mild orthokeratosis, papillomatosis, heavily pigmented epidermis, liquefaction degeneration of the basal layer, dermal pigmentary incontinence, melanophages, interface dermatitis, and sparse, superficial perivascular inflammations.[6] The microscopic examination of the hair shaft showed normal hair shafts [7] as observed in our patients.Although DPR and NFJS have poorly developed dermatoglyphics, specifically reticulate hyperpigmentation of the skin, DPR has been distinguished from NFJS by the lifelong persistence of the skin hyperpigmentation, partial alopecia, and absence of dental anomalies.[8]

No specific laboratory changes are seen in DPR. The typical histopathologic picture of DPR shows liquefaction degeneration of the basal layer and dermal pigmentary incontinence. No specific treatment exists for DP, except for symptomatic management of some of the associated conditions, such as palmoplantar hyperkeratosis. For hyperkeratosis, topical retinoic acids and keratolytics may be tried.

How to cite this article:
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CASE REPORT
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WERNER'S SYNDROME: A RARE ENTITY

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Abstract

Werner syndrome is a rare inherited adult premature ageing syndrome in which the ageing process is accelerated after puberty. Clinically characterized by short stature, scleroderma-like skin alterations, cataracts and premature ageing of the face. We report a case of this rare entity fulfilling three major and two additional signs.

Key words: Werner, scleroderma, cataract

Introduction

Werner syndrome (WS) also known as pangeria or progeria adultorum is an inherited adult premature ageing syndrome in which the ageing process is accelerated after puberty. It is an autosomal recessive disease and observed more commonly with family history of consanguinous marriage. Its clinical manifestations include short stature, scleroderma-like skin alterations, cataracts and premature ageing of the face.[3]

Case Report

A 45 year old male patient presented to the medicine department with complaints of fever with palpitations since 1 month and there was history of breathlessness on exertion since 10 days along with pain abdomen and joint pains. He was referred to dermatology department for baldness since adolescence, reduced body hair and dryness of skin. On further enquiry there was family history of consanguinity and similar complaints in father and older male sibling.

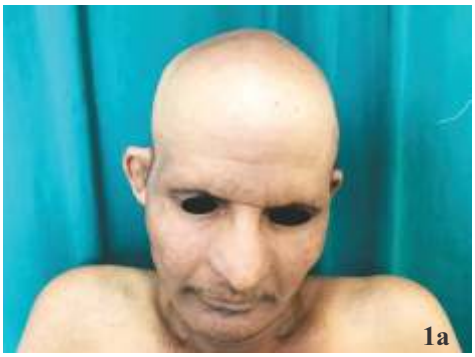
On clinical examination patient weighed 42kg with short stature and slender extremities. Scalp showed diffuse non scarring alopecia involving most of the scalp (sparing occipital hair) (fig1a) with loss of body hair including axillary and pubic hair (fig 1b), sparse eyebrows, greying of beard hair, beak like nose (bird like facies) , peg shaped tooth (fig 2), atrophy of the skin, poikilodermatous changes over the upper back (fig 3), ichthyotic changes over the lower back with syndactyly involving both hands and feet (fig 4a,b), flat feet, clubbing of finger nails and a high-pitched voice.

Blood work up revealed low haemoglobin (8.9g%) and raised levels of thyroid stimulating hormone (TSH- 14.16 IU), liver function tests showed raised total bilirubin (2.5mg/dl) with normal liver enzymes and high fasting blood sugar levels(210mg/dl) .Cardiovascular assessment showed normal blood pressure and echocardiography revealed severe mitral stenosis, grade2 mitral regurgitation, grade1 aortic regurgitation, mild tricuspid regurgitation suggesting rheumatic heart disease. Ultrasound of abdomen and pelvis showed splenomegaly. Radiograph of the limbs showed no osteoporosis. Skin biopsy showed loss of adnexal structures in the dermis. Ophthalmalic examination could not be done.

Discussion

Progeroid syndromes are a heterogeneous group of disorders with variable cutaneous features that lead to premature ageing, including poikiloderma, photosensitivity, sclerodermatous changes, alteration of the subcutaneous fat, or skin laxity and wrinkling.

Werner syndrome is a rare, autosomal recessive disorder caused by mutations in the gene RECQL2 (WRN) on chromosome 8p12-p11.2 which encodes for DNA helicase. Aberrant repair of double-stranded DNA damage in the absence of WRN helicase activity leads to an accumulation of DNA damage, telomere shortening, genetic instability and a reduction in cellular replicative lifespan. The tissues of mesenchymal origin are affected preferentially compared to neural tissues.[5]



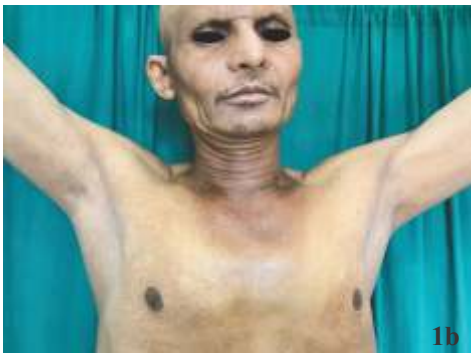
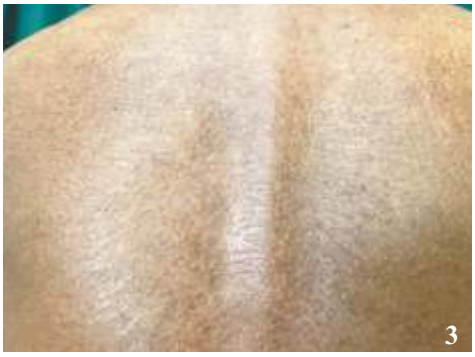
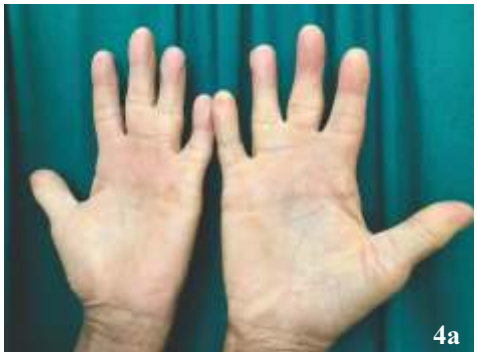


Figure 1 (a,b) & 2 : 1a. non sacrring alopecia of scalp. **1b.** loss of axillary hair. **2.** beaking of the nose with peg shaped tooth





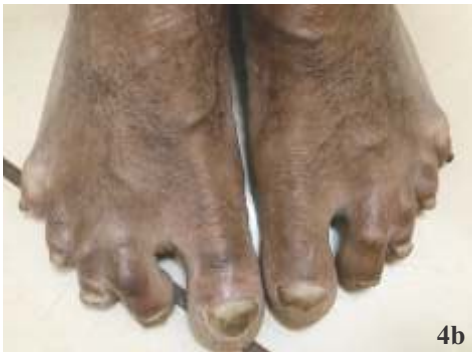


Figure 3 & 4 (a,b) : **3.** poikiloderma of upper back. **4 (a,b).** syndactyly of hands and feet

The prevalence of this genetic syndrome varies with the rate of consanguinous marriage in the population and estimated incidence is 1 case in 1million individuals. It is more prevalent in Japanese population 1/20,000 to 1/40,000 and in the U.S. population estimated prevalence is 1/20,000.[1]

Originally this syndrome was first described by Otto Werner in 1904. He noted the following clinical features: short stature, scleroderma-like skin alterations, cataract, premature aging of the face, grey hair and genital hypoplasia in 4 siblings. Oppenheimer and Kugel in 1934 reported the presence of additional endocrinological abnormalities such as osteoporosis and hyperglycemia.[3]

Patients with WS usually develop normally until the third decade of life. Usually, the first clinical sign is a lack of the pubertal growth spurt during the

teenage. In the second and third decade of life these patients begin to manifest with skin changes like atrophy, loss of hair and graying of hair. Some patients may present with a high-pitched voice and flat feet.[4]

Subsequently WS patients develop common age related changes like type2 diabetes mellitus, atherosclerosis, osteoporosis and malignancies. The average life expectancy is around 50 years. The studies conducted by Epstein et al showed that the common cause of death was malignancy and myocardial infarction.[4]

Mesenchymal sarcoma is seen 10 times more common.Other malignancies with elevated incidences are malignant melanoma, thyroid cancer, osteosarcoma, and soft tissue sarcoma. Immunological and DNA abnormalities are found to be associated with development of malignancies.[2]

Since WS has an autosomal recessive trait, established cases of WS should be referred for genetic counseling to ensure early identifcation and treatment of syndrome-associated manifestations.

For clinical assessment of WS a diagnostic criteria is used. It was originally proposed by Nakura et al in 1994 (Table 1). A definitive diagnosis is made when all major signs and two additional signs are present. When first three major signs and any two other signs are present probable diagnosis is made. If either cataracts or dermatological alterations and any four other signs are seen then a possible diagnosis of WS is made.[4]

The above reported case fulfils three of the major criteria with two additional signs hence a probable diagnosis of WS was made.

Major criteria: 1. Cataracts (bilateral) 2. Characteristic dermatological pathology (tight skin, atrophic skin, pigmentary alterations, ulceration, hyperkeratosis, regional subcutaneous atrophy) and characteristic facies ('bird-like' face) 3. Short stature 4. Premature greying and/or thinning of scalp hair [Parental consanguinity (third cousin or closer) or aVected sibling] [Positive 24-h urinary hyaluronic acid test when available]
Additional signs and symptoms: 1. Type 2 diabetes mellitus 2. Hypogonadism (secondary sexual underdevelopment, diminished fertility, testicular or ovarian atrophy) 3. Osteoporosis 4. Osteosclerosis of distal phalanges of fingers and/ or toes (x-ray diagnosis) 5. Soft tissue calcification 6. Evidence of premature atherosclerosis (e.g., history of myocardial infarction) 7. Neoplasms: mesenchymal (i.e. sarcomas), rare (unusual), or multiple 8. Abnormal voice (high-pitched, squeaky, or hoarse) 9. Flat feet
Definite diagnosis: All the major signs and two additional signs Probable diagnosis: The first three major signs and any two others signs Possible diagnosis: Either cataract or dermatological alterations and any four other signs Exclusion of diagnosis: Onset of signs and symptoms before adolescence (except short stature)

Table 1: Clinical diagnostic criteria[4]

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CASE REPORT
Year: 2019 • Volume: 2 • Issue: 1 • Page: 26-27

GENERALIZED MORPHEA IN A CHILD – A RARE ENTITY

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Abstract

Generalized morphea is a rare presentation, if present in children. We present a 5-year old female child who developed tautness of skin since 3 months.

Key words: generalized morphea, children, rare

Introduction

The term "Morphea" encompasses a group of related conditions characterized by varying degrees of sclerosis, fibrosis and atrophy in skin and subcutaneous tissue, sometimes extending deeply into muscle, bone and brain.[1] Anti-nuclear antibody(ANA) positivity is common but the specific autoantibodies seen in systemic sclerosis are rarely present.[1] Various type of morphea are – Limited morphea, Generalized morphea , Linear morphea & Mixed type. Generalized morphea is a rare condition, it occurs in the cases of dissesminated sclerosis with no systemic involvement. It occurs mainly in adults.It begins in the trunk and causes contractures and deformities in limbs. It differentiates from systemic sclerosis because it does not accompany Raynaud's phenomenon and doesn't show capillaroscopic changes.[2] The disease is extremely uncommon in children.[3]

Case Report

A 5-year female child presented with gradual onset and progressive tautness and tightness of skin since 3 months, which was accompanied by limitation of movement of hands,elbow,ankle and knee joints. Patient also complained of pain over fingers and toes without any change in colour. There was no history of regurgitation of food and difficulty in breathing. There was no history of trauma,bites or history of inflammation over those sites On examination, multiple large shiny atrophic plaques of size ranging from 3 cm to 20 cm were present over bilateral upper limb and lower limb involving the joints [Figure 1 and 2].

Contracture of small joints of hand was present, limiting the movement of hand [Figure 3].

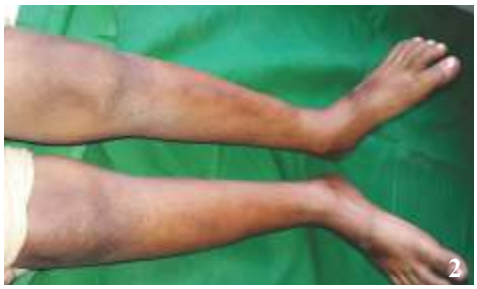
Matt telangiectesia was seen over lips with telangiectesia over bilateral cheeks(without any history of any topical application)[Figure 4 and 5].

Other dermatological examination was unremarkable. Based on history and clinical examination, a differential diagnosis of generalized morphea and CREST syndrome was considered. Complete blood count, ESR thyroid profile, Anti-nuclear antibody(ANA), Extractable nuclear antibody(ENA) profile and upper GI endoscopy were all within normal limit.Histopathology revealed dermal stroma with packed collagen bundles, eccrine glands with replacement of periadnexal fat by collagen and moderate lymphocytes, plasma cells and histiocytes around vessels [Figure 6 and 7].

Based on clinicohistological correlation,a diagnosis of generalized morphea was made. Patient was started with oral prednisolone (10 mg),topical calcipotriol and physiotherapy with an improvement of around 40%.

Discussion

When scleroderma of any type affects children, it is called Juvenile Scleroderma.[4] They constitute about 10% of patients suffering from scleroderma.[5] Generalized morphea is rare in infants. The diagnostic criteria of generalized morphea consists of – 1. 4 or more plaques become larger than 3 cm and merge, 2.Involvement of 2 or more anatomical areas [Head and neck, left or right upper limb or lower limb, anterior and posterior



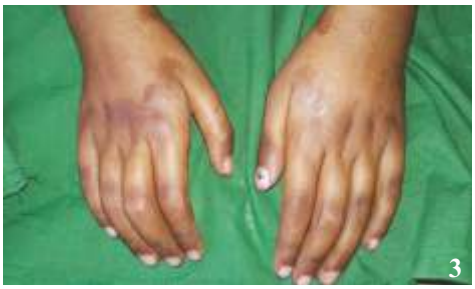


Figure 1,2, 3 : **1**. Large plaques of morphea over right upper limb. **2**. Large plaques of morphea over both legs. **3**. Contracture of small joints of hands

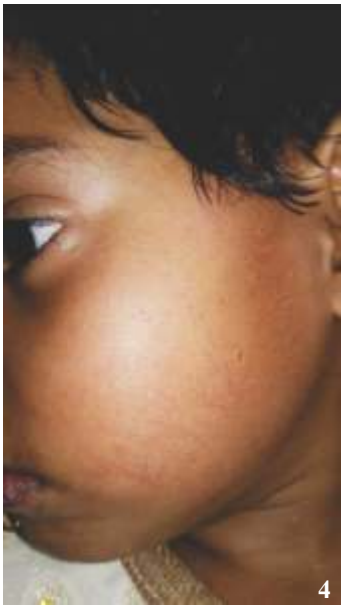
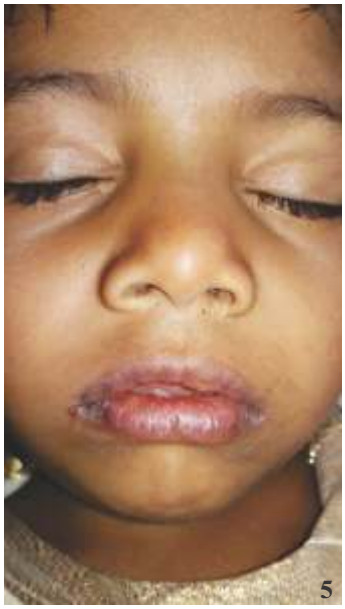



Figure 4, 5 : **4**. Telanigectesia over face. **5**. Matt telangiectesia over lips

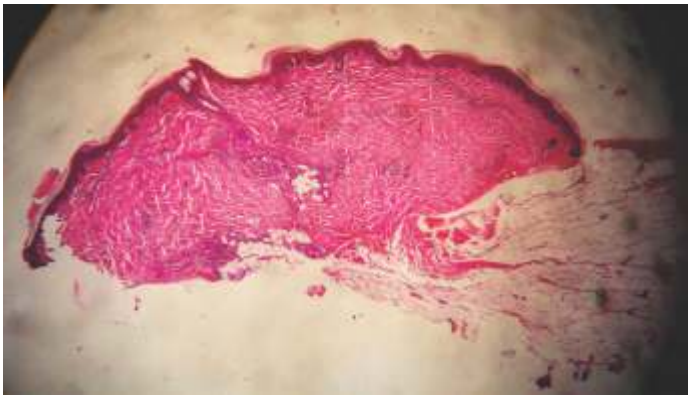


Figure 6 : Epidermal atrophy and dermal stroma with thick collagen bundles. (H & E Scanner View)

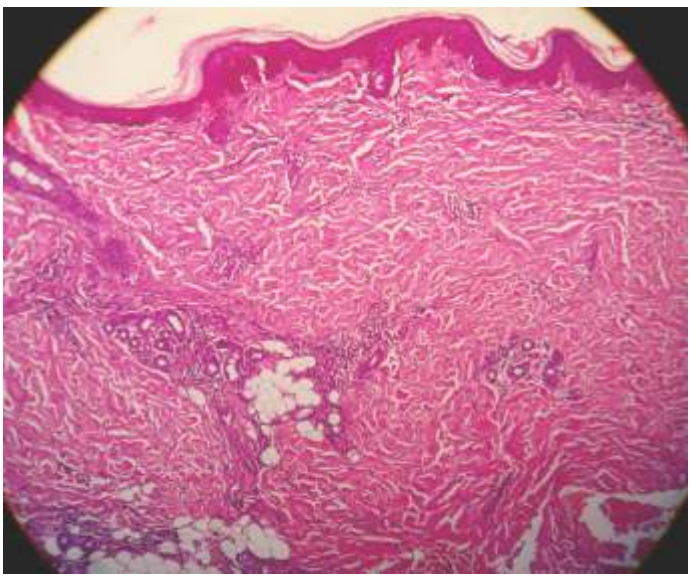


Figure 7 : Dermal stroma with thick collagen bundles, atrophy of pilosebaceous units with hypertrophied piloarrector muscle, presence of eccrine sweat glands in the dermis with replacement of periadnexal fat by collagen and moderate lymphocytes, plasma cells and histiocytes around blood vessels. (H & E 10×)

trunk].[6] The diagnosis of morphea is primarily clinical.

We excluded the differential of CREST syndrome by means of history and negative Anti-nuclear antibody and Extractable nuclear antibody profile

In a large multicentre, international study of Juvenile Localized Scleroderma of 750 children, generalized morphea was present in 7% of patients. It is rare during childhood.[7]

In 1976,Ansell et al[8] described 2 cases (10-year girl and 8-year boy) of generalized morphea with fixed flexion deformities of joints at the site of morphea lesions, similar to our case.

In 2003,Brar BK et al[3] reported a 9-year girl with generalized morphea, following measles vaccination.

In 2015,PK Dey et al[9] described a 10-month male infant with generalized morphea and reported its response to topical steroids.

The above cases lacked presence of telangiectesia over lips and face which was present in our case and drifted us to rule out CREST syndrome.

Only a handful cases have been reported from the Indian subcontinent, hence this case is being reported.

How to cite this article:
Jain S, Mohapatra L, Mohanty P, Sebastian E.Generalized Morphea In A Child – A Rare Entity. JDA Indian Journal of Clinical Dermatology 2019;2:26-27.

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QUIZ

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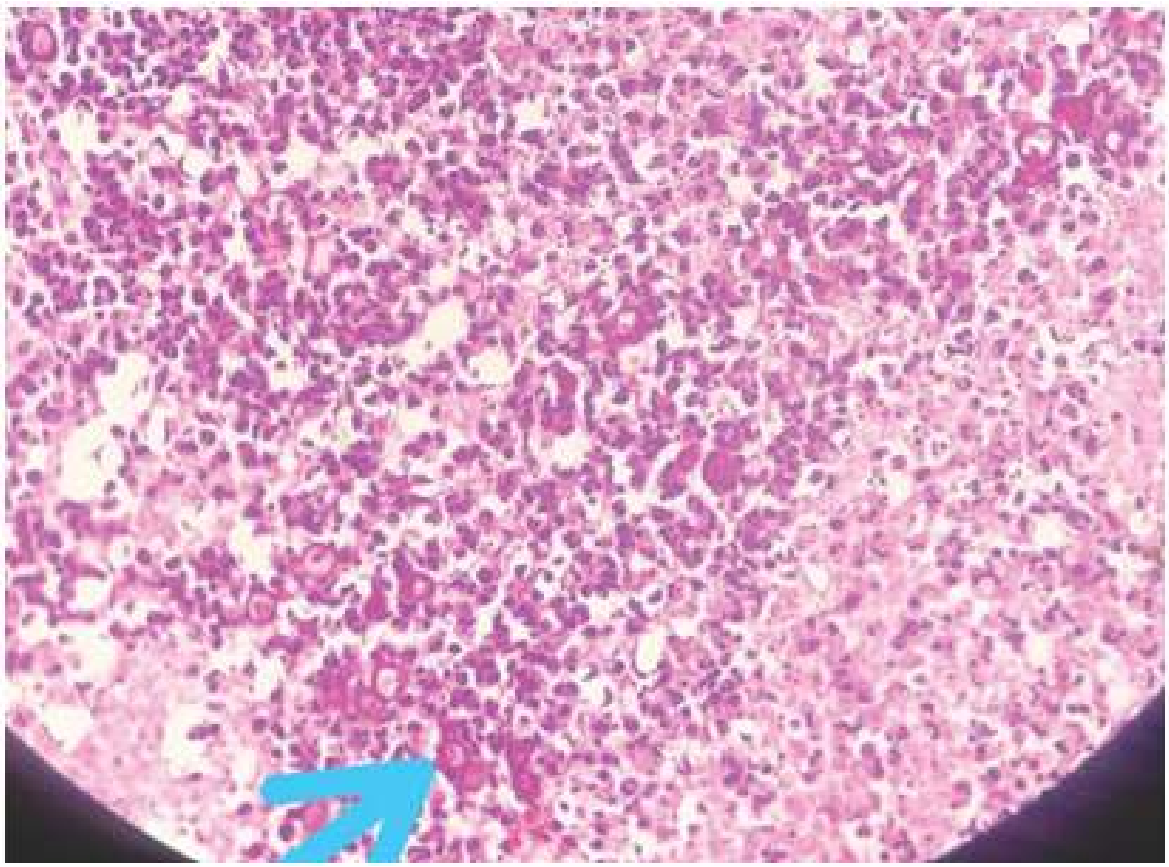
A 42-year old otherwise healthy male farmer presented with a slowly spreading, small, hyperpigmented, moderately itchy, irregular shaped plaque with warty surface of 3 * 5cmsin size approximately of about 4months duration present over left leg laterally just above lateral malleoli. Dermatological examination revealed single round to oval, well defined verrucous plaque located on the lateral aspect of the lower part of his left leg. Patient couldn't recall any history of trauma.

His physical and systemic examination were unremarkable andhematological and biochemical laboratory parameters were within normal limits. Radiological examination did not reveal any bone involvement. Biopsy revealed hyperkeratosis, acanthosis, and pseudoepitheliomatous hyperplasia, intraepidermalabcess. The reticular dermis showed granulomatous inflammatory cell infiltrate with giant cells containing several rounded, brownish (copper-colored), thick-walled structures (fig.-1). No mycelial elements were seen.

What is your diagnosis?



(For answer visit PG Quiz section at **www.e-ijcd.in**)

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