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Dr. Sori Tukaram
Assistant Professor,
Department of Skin & STD,
GIMS, Gadag, Karnataka,
India

Dr. Dyavannavar Veeresh V
Associate Professor,
Department of Skin & STD,
GIMS, Gadag, Karnataka,
India

Dr. TJ Jaisankar
Professor, Department of Skin
& STD, GIMS, JIPMER,
Pondicherry, India

Dr. Thappa DM
Professor, Department of Skin
& STD, GIMS, JIPMER,
Pondicherry, India

Corresponding Author:
Dr. Dyavannavar Veeresh V
Associate Professor,
Department of Skin & STD,
GIMS, Gadag, Karnataka,
India

Original Article

A study of dermoscopy for pigmentary lesions among children with pigmentary disorders

Dr. Sori Tukaram, Dr. Dyavannavar Veeresh V, Dr. TJ Jaisankar and Dr. Thappa DM

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Abstract

Disorders of melanin pigmentation can be divided on morphological grounds into two types- hypomelanosis and hypermelanosis. Hypomelanosis is characterized by a lack of melanin pigment in the skin, which therefore appears white or lighter than the normal colour. In hypermelanosis, there is an increased amount of melanin in the skin. When this excess is confined to the epidermis, the skin appears browner than normal, and when present in the dermis, a slate-grey or blue appearance is produced. Hypermelanosis and hypomelanosis can be generalized and diffuse, or may be localized and circumscribed. Amelanosis is the term applied when there is a total lack of melanin in the skin. A detailed clinical history was elicited with regard to patients' age, gender, address, age of onset of disorder, type (hypo-/hyper pigmented), site and size of pigmented lesions, familial involvement, associated skin and systemic conditions and the details were recorded on a proforma. A dermoscope 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. Basically, a dermoscope is functionally similar to a magnifying lens but with the added features of an inbuilt illuminating system with the ability to assess structures as deep as in the reticular dermis, and the ability to record images.

Keywords: Dermoscopy, pigmentary lesions, children

Introduction

Melanin is a pigment produced by melanocytes, which are specialized dendritic cells derived from the neural crest that migrate to the basal layer of the epidermis during embryogenesis. Melanocytes synthesize and package melanin within discrete membrane-bound organelles called melanosomes, which are then transferred via melanocytic dendrites to the surrounding keratinocytes of epidermis and hair follicles. Dark individuals have more numerous, larger, singly dispersed melanosomes, whereas individuals with light complexion have fewer, smaller melanosomes that are aggregated into complexes and are degraded more rapidly. The presence of melanin in the epidermis helps to protect against ultraviolet radiation and associated cutaneous damage, including pigmented nevi, actinic damage, and possible cutaneous neoplasia [1].

Disorders of melanin pigmentation can be divided on morphological grounds into two types- hypomelanosis and hypermelanosis. Hypomelanosis is characterized by a lack of melanin pigment in the skin, which therefore appears white or lighter than the normal colour. In hypermelanosis, there is an increased amount of melanin in the skin. When this excess is confined to the epidermis, the skin appears browner than normal, and when present in the dermis, a slate-grey or blue appearance is produced. Hypermelanosis and hypomelanosis can be generalized and diffuse, or may be localized and circumscribed. Amelanosis is the term applied when there is a total lack of melanin in the skin. Alterations in pigmentation can occur due to a variety of genetic and environmental factors.

Freckles are well-circumscribed macules, usually 2-3mm in diameter, reddish-tan and brown in color, which appear in childhood and tend to fade in adult life. They commonly arise, in childhood, especially between 2 and 4 years of age, and their presence correlates with fair skin, red hair, and an increased risk of developing melanoma. Freckles are most common on the sun-exposed areas of the face, shoulder and upper back. Lesions tend to fade in the winter

and increase in number during spring and summer months. Freckles bear cosmetic, but no systemic significance. Freckles appear as a result of functionally overactive melanocytes (though they are normal in number). Histology reveals excess of melanin pigment in the basal layer^[2].

Becker's nevus is a relatively common anomaly, and is about five times more frequent in males than in females and characterized by solitary large hyperpigmented macule or multiple small coalescing macules in a geographical contour, over the shoulder, anterior chest, or scapular region. Thickening of skin over the centre of the lesion, and appearance of thick, dark hairs on and around the lesion can be seen more frequently. Becker's nevus (Becker's melanosis) is sometimes congenital, but more commonly starts during late childhood or childhood or early adolescence. The etiology of Becker's nevus is unknown, but a localized increase in androgen receptor sensitivity may explain the time of onset and clinical features seen in most individuals with this disorder^[3].

Association of a variety of non-cutaneous abnormalities has been described, especially unilateral hypoplasia of the breast in the females. Aplasia of the ipsilateral pectoralis major muscle, ipsilateral limb shortening, localized lipodystrophy, spina bifida, scoliosis, pectus carinatum, congenital adrenal hyperplasia and an accessory scrotum had also been found to be associated (Becker's nevus syndrome). Multiple Becker's nevi and bilateral Becker's nevi has been reported.

Lentigines are small, tan, dark brown or black flat, oval, or circular, sharply circumscribed lesions that usually appear in childhood and may increase in number until adult life. Each lesion ("lentigo simplex") usually measures 3-15 mm in diameter and may occur on any cutaneous surface. The pigmentation is uniform and darker than that seen in ephelides (freckles) and café-au-lait macules, and the color is unaffected by exposure to sunlight. Lentigines are typically larger than freckles and smaller than a typical café-au-lait macule. The major lentiginous syndromes are Peutz-Jeghers, multiple lentiginosis/LEOPARD syndrome, and Carney complex.

Peutz and Jeghers initially described this autosomal dominant disorder of mucocutaneous lentiginous macules and multiple hamartomatous intestinal polyps in 1921 and 1949, respectively. PJS is a rare disease. The incidence is 1 in 30,000 to 1,20,000 live births. The disease affects males and females equally.

PHS is characterized by bluish brown to black spots, often present at birth or in early childhood, which represent a cutaneous marker of this syndrome. These discrete, flat pigmented lesions are irregularly oval, and usually measure less than 5 mm in diameter. They are mostly seen on the lips and buccal mucosa, nasal and periorbital regions, elbows, dorsal aspects of the fingers and toes, palms, soles, and periumbilical, perianal, or labial regions; occasionally the gums and the hard palate and, on rare occasions, even the tongue may be involved. Although the polyps of Peutz-Jeghers syndrome are generally benign, adenocarcinomas of gastrointestinal tract (stomach, small intestine, colorectal, pancreas, and biliary tract), breast and uterine cervical carcinomas, and gonadal sex tumours (Sertoli cell tumours of the testis and tumours of the ovary) have been described.⁴ Most patients have a characteristic clinical course of recurrent episodes of polyp induced bowel obstruction and bleeding. At pediatric age, major clinical impact is related to complications associated to intestinal polyps, but neoplastic

risk isn't negligible. Though clinical surveillance is recommended since the age of 10 years, relevant lesions may occur before that age.

Café-au-lait spots are large, round or oval, flat lesions of light brown pigmentation found in up to 10- 33% of normal children. Frequently present at birth, or developing soon thereafter, vary from 1.5 cm or less in their smallest diameter to much larger lesions that may measure up to 15 to 20 cm or more in diameter. Although most individuals with café-au-lait spots are normal, these pigmented macules may be a sign of neurofibromatosis and other systemic disorders.

Café-au-lait spots may be present on any cutaneous surface at birth or may appear later in life. With about 10% of the normal individuals having one to three of these macules, it is not surprising that the café-au-lait spots have been reported in a number of syndromes. Three disorders in which café-au-lait spots are especially prominent in size and number are neurofibromatosis, McCune-Albright's syndrome, and Watson's syndrome.

Neurofibromatosis is an autosomal dominant disorder characterized by an increased propensity towards the development of tumours of the nerve sheath. Neurofibromatosis encompasses two distinct disorders, neurofibromatosis type 1 (NF-1) which occurs in 1 in 3500 births and neurofibromatosis type 2 (NF-2) which occurs in 1 in 40,000 births. Café-au-lait spots and dermal and plexiform neurofibromas are the characteristic cutaneous findings of NF-1^[2].

A dermoscope (Syn: dermatoscope, skin surface microscope, epiluminescence microscope or episcope) 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 advantages of this procedure are: it obviates the need for a skin biopsy, the images are available immediately and can be easily stored^[2]. Basically, a dermoscope is functionally similar to a magnifying lens but with the added features of an inbuilt illuminating system with the ability to assess structures as deep as in the reticular dermis, and the ability to record images.

Methodology

Children (up to 14 years of age) with pigmentary disorders were included in the study after getting informed consent from the parents/guardians. Children more than 14 years of age, and when parents/guardians (of children less than 14 years of age) are not consenting for participation in the study were excluded.

A detailed clinical history was elicited with regard to patients' age, gender, address, age of onset of disorder, type (hypo-/hyper pigmented), site and size of pigmented lesions, familial involvement, associated skin and systemic conditions and the details were recorded on a proforma. Detailed examination of the pigmentary lesion/lesions was done and findings were noted in terms of site, size, shape, morphology of lesions etc. General physical examination was done to see any associated cutaneous or systemic involvement and the relevant findings were recorded.

Dermoscopy was done for the patients, where the pigmentary lesions were difficult to diagnose and characterize clinically. Dermoscopy was done with Hanse HVS-CM500P (USB Type Connectable to Computer) using contact technique the glass plate of the instrument in direct

contact with the surface of the linkage fluid applied lesion (liquid paraffin was the linkage fluid used, which was inexpensive, safe, and easily available, with good results); glass slide was placed over the oil applied lesion, as a contact plate. As glass has a refractive index of 1.52, which is similar to that of skin (1.55), it further enhances transillumination of the lesion. Images of dermoscopy were stored in a computer

(software provided by the Hanse Electronic Co. Ltd). Skin biopsy was done as and when indicated. Photographs were taken in patients who agreed for that. Results were tabulated and analyzed using SPSS 13.0 software.

Results

Table 1: Age-wise distribution of the pigmentary disorders seen in our study

Diagnosis		Number of children				Total
		<1 year	1-5 years	6-10 years	11-14 years	
Hyperpigmentary disorders	Café au lait macule	1	6	1	4	12
	Post-inflammatory hyperpigmentation	1	2	3	3	9
	Pigmentary mosaicism	4	2	1	1	8
	Congenital melanocytic nevus	1	2	2	2	7
	Lichen planus	-	-	3	2	5
	Mongolian spots	2	1	-	1	4
	Fixed drug eruption	1	2	-	-	3
	Becker's nevus	-	-	-	2	2
	Urticaria pigmentosa	1	-	1	-	2
	Chediak-Higashi syndrome	-	1	-	-	1
	Discoid lupus erythematosus	-	-	-	1	1
	Incontinentia pigmenti	1	-	-	-	1
	Centrofacial lentiginosis	-	-	-	1	1
	Lichen striatus	-	-	1	-	1
	Nevus of Ota	-	-	-	1	1
	Pityriasis rubra pilaris	-	-	1	-	1
Speckled lentiginous nevus	-	-	-	1	1	
Hypopigmentary disorders	Pityriasis alba	3	11	8	6	28
	Vitiligo	-	4	6	13	23
	Leprosy	-	1	4	8	13
	Nevus depigmentosus	2	2	6	1	11
	Tinea versicolor	2	2	1	2	7
	Hypomelanosis of Ito	1	2	-	2	5
	Post-inflammatory hypopigmentation	0	2	2	1	5
	Pityriasis rosea	-	3	1	-	4
	Steroid induced hypopigmentation	3	1	-	-	4
	Lichen sclerosus et atrophicus	-	-	2	1	3
	Pityriasis lichenoides chronic	-	-	3	-	3
	Lichen striatus	-	2	-	-	2
	Oculocutaneous albinism	-	1	1	-	2
	Tuberous sclerosis complex	-	-	1	1	2
	Hypopigmentary mosaicism	1	-	-	-	1
Griscelli syndrome	-	-	1	-	1	
Dyschromatosis universalis hereditaria		-	-	-	1	1

Table 2: Systemic involvement/syndromic associations in hyperpigmentary disorders in children

Pigmentary lesions	Age & Sex	Eyes	Oral mucosa	CVS/Echocardiography	RS	CNS	Visceral involvement	Musculo-skeletal	Syndromic Diagnosis
Mongolian spots	1 yr F	Corneal haziness	Normal	Mild left ventricular hypertrophy	Normal	Wide anterior fontanelle with enlarged calvarium on X-ray skull	Hepato-splenomegaly	Normal	Mucopolysaccharidosis
Mongolian spots	9 mth F	Corneal haziness	Normal	Left atrial and left with moderate to severe mitral regurgitation with mild aortic prolapse	Normal	Normal	Hepato-splenomegaly	Normal	Hurler's syndrome
Centrofacial lentiginosis	11 yr M	Hypertelorism	Normal	Normal	Normal	Normal	Normal	Normal	-
Diffuse hyperpigmentation	2 yr F	Normal	Normal	Normal	Recurrent respiratory infections	Delayed milestones	Hepato-splenomegaly	Normal	CHS
CALM	5 yr M	Normal	Normal	Normal	Normal	Seizures	Normal	Normal	TSC
Bizarre, splashed, linear	1 yr M	Esotropia	Abnormal dentition	Normal	Normal	Spastic diplegia	Normal	Normal	Incontinentia pigmenti
CMN	4 yr M	Normal	Normal	Normal	Normal	Normal	Normal	Limb length discrepancy	CMN
CALM	10 yr F	Normal	Normal	Normal	Normal	Normal	Normal	Short stature	McCune-Albright syndrome

Discussion

A dermoscope 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. Basically, a dermoscope is functionally similar to a magnifying lens but with the added features of an inbuilt illuminating system with the ability to assess structures as deep as in the reticular dermis, and the ability to record images.

Café-au-lait macule showed following Dermatoscopy structures in our study typical network and reticular pattern. The pigment network is a grid-like (honeycomb-like) network consisting of pigmented “lines” and hypopigmented “holes”. The reticulation (network) represents the rete ridge pattern of the epidermis. Its histopathological correlation is either melanin pigment in keratinocytes, or in melanocytes along the dermo-epidermal junction. The hypopigmented holes in the network correspond to tips of the dermal papillae and the overlying supra-papillary plates of the epidermis. Café-au-lait macule is melanotic disorder, in which there is increased amount of melanin in the basal layer, which would explain the dermoscopic features in our children. To the best of our knowledge, dermoscopic features of CALM have not yet been described in the literature. The pigment networks noted in our patients were relatively uniform, regularly meshed, and homogeneous in color with thinning out at periphery^[5]. In a study by Ingordo *et al*, the dermoscopic features of 127 CMN were pigment network (114), follicles (86), vessels (84), focal hypopigmentation (68), skin furrow hypopigmentation (62), globules (54), focal thickening of network lines/network (52), target network (36), blotches (25), homogenous diffuse pigmentation (22), target globules (21), target vessels (12), hyperpigmented areas (3), perifollicular hypopigmentation/follicles (29/45). The dermoscopic features observed in our CMN patients were typical pigment network, reticular pattern and globular pattern (which is diffuse throughout), and homogenous pattern.

LP can demonstrate the following dermoscopic features: diffuse structureless, brownish areas, fine or coarse gray-blue or brown dots or globules, and Whickam striae (polymorphic pearly whitish structures) which are pathognomonic of lichen planus. Vázquez-López *et al* studied mature, active violaceous papules and plaques of LP and noted characteristic rounded, arboriform, reticular, or annular Whickam striae (WS) in dermoscopy. Dermoscopic features observed in our lichen planus patients were blue-white structure (5), WS (4), globules at center (1), radial migration lines at periphery (1), dots at periphery (1), structureless areas (1), coarse gray-blue globules (1), and dotted vessels (1). WS was not observed in one case of LP. Dermoscopic features of psoriasis are mainly homogenous red dots with some patches of pink to bright red (dotted vessels)^[6].

In a study by Vázquez-López *et al*, dermoscopy in psoriasis showed dilated red globules which were regularly arranged and uniformly distributed along the psoriatic plaque. In a subpattern of plaque-type of psoriasis, the red globules (presence of round capillaries) were arranged in irregular circle or rings with a beaded, lacelike capillary appearance. Histologically, red globules represent the top of the tortuous, plentiful, coiled, ectatic, and elongated capillaries

within the thin, elongated, psoriatic papillae, which are the vascular basis of the Auspitz sign^[7].

In our study, we observed dermoscopy features of psoriasis in children (mainly on the subsiding lesion) were uniform distribution of red dots, dotted vessels on pink homogenous background, White structureless areas, dotted vessels, pink homogenous area, White structureless areas with uniform distribution of red dots.

Dermoscopic features observed in our Becker’s nevus patients were, typical pigment network (2) with diffuse homogenous thickening and reticular pattern (2) in two cases on BN^[8].

In a study by Ingordo *et al*, dermoscopic features of 64 cases of BN were pigment network (62), vessels (46), follicles (45), focal hypopigmentation (30), skin furrow hypopigmentation (25), target network (13), and globules (8). Pigment network was in the form of focal thickening of network lines/network (9/62), blotches (25), homogenous diffuse pigmentation (1), target globules (1), and perifollicular hypopigmentation/follicles (29/45)^[9, 10].

Fixed Drug Eruptions

The dermoscopic features of fixed drug eruption in our study were typical pigment network, and multiple small black dots.

Tuberous Sclerosis Complex (TSC)

Dermoscopy of angiofibroma showed, yellowish-brown oval structures with branching vessels inside the globules. Dermoscopy of forehead plaque and ash-leaf macule showed milia-like cyst and irregular distributed structureless areas.

Urticaria Pigmentosa

The dermoscopic features of urticaria pigmentosa in our study was typical pigment network, reticular pattern.

Pigmentary Mosaicism

The dermoscopic features observed in our pigmentary mosaicism patients were typical pigment network, reticular pattern, and milia-like cyst.

Linear and Whorled Hyperpigmentation

The dermoscopic features of linear and whorled hyperpigmentation in our patients were linear or circular arrangement of streak like pigmentation with gyriform structures. In a study by Ertam *et al*, dermoscopic examination showed linear or circular arrangement of streak-like pigmentation arranged in a parallel manner.

Linear Nevoid Pigmentation

In our study, linear nevoid pigmentation showed reddish brown blotches.

Speckled Lentiginous Nevus

Speckled lentiginous nevus in our study showed globular structure with reddish brown blotches.

Pityriasis Rubra Pilaris

The dermoscopic features observed in pityriasis rubra pilaris were asymmetrical follicular round to ovoid structures and blue-white structures.

Hypomelanosis of Ito

The dermoscopic features observed in hypomelanosis of Ito were structureless areas, milia-like cyst, and streak like pigmentation.

Hypopigmentary Mosaicism

Hypopigmentary mosaicism in our study showed structureless areas with network pattern.

Lichen Striatus

The dermoscopic features observed in lichen striatus patient were structureless areas and milia-like cyst.

Vitiligo

The dermoscopic features observed in our vitiligo patient were structureless areas with brown dots.

To conclude, a wide range of pigmentary disorders are common in children. Both hyper- and hypopigmented disorders are seen in children; but, as is evident from our study, hypopigmentary disorders appear to be commoner in children. The most common pigmentary disorder in our study was pityriasis alba (28/167, 16.7%) followed by vitiligo (23/167, 13.7%), leprosy (13/167, 7.78%), café-au-lait macule (12/167, 7.18%), nevus depigmentosus (11/167, 6.58%), postinflammatory hyperpigmentation (9/167, 5.38%), pigmentary mosaicism (8/167, 4.79%), congenital melanocytic nevus (7/167, 4.19%) and tinea versicolor (7/167, 4.19%). While most of the pigmentary disorders in our study conformed to the characteristics already described in the literature, some of our cases had rather unique features hitherto unreported in the literature such as the pattern of lesions in the male child with incontinentia pigmenti, pattern of phylloid pigmentary mosaicism on the face, diffuse hyperpigmentation in the child with Chédiak-Higashi syndrome and tinea versicolor infection involving the medial canthi of eyes in infants. Dermoscopy is useful tool to confirm the diagnosis in pigmentary disorders.

Conclusion

The dermoscopic features were, blue-white veil, and globules at centre with radial migration lines at periphery in lichen planus, asymmetrical follicular round to ovoid structures in pityriasis rubra pilaris, milia-like cyst in forehead plaque and yellowish-brown ovoid structures with branching vessels inside the structure.

Dermoscopy is an important instrument which can help in diagnosis of congenital melanocytic nevi, Becker's nevus, lichen planus, psoriasis and other pigmentary disorders without need of biopsy, especially in children. The images can be stored and follow up of the patients can be done to know any improvement or any malignant changes especially in cases of congenital melanocytic nevi.

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