Ashy dermatosis in an 8-year-old Indian child

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ABSTRACT

Ashy dermatosis is a disorder of pigmentation, characterized by asymptomatic symmetric ashy gray-colored macules, in the first to third decade of life. It can, however, affect children sometime. But, there is a paucity of similar cases in Indian children. We present a case of ashy dermatosis in an 8-year-old Indian girl who presented to skin OPD with areas of ashy pigmentation, which were distributed symmetrically. Skin biopsy was consistent with lichenoid pattern histology. This case has been reported to highlight the rarity of ashy dermatosis in Indian children.

Key words: Ashy dermatosis, children, erythema dyschromicum perstans

INTRODUCTION

Ashy dermatosis or EDP is an uncommon entity, which occurs worldwide in all races and is characterized by symmetric ashy gray-colored confluent macules with polycyclic margins over the body. The etiology of ashy dermatosis or EDP is unknown. However, a number of etiological factors have been implicated.

CASE REPORT

An 8-year-old dark-skinned female child of Indian origin presented to the dermatology outpatient department with asymptomatic, gradually progressive hyperpigmented flat lesions over the trunk of 1-year's duration. There was no history of mustard oil application or use of drugs prior to the onset of lesions. There was no complaint of pruritus, fatigue, or symptoms suggestive of any systemic disease.

The general and systemic examination did not reveal any abnormality. Cutaneous examination revealed ashy gray-colored, confluent, symmetrical macules with polycyclic margins over almost the entire trunk, proximal upper limb, posterior neck, proximal thigh, forehead, and left upper eyelid [Figure 1a and b]. Only a few lesions could be discerned as isolated lesions, which were variable in size, from 0.5 cm to 2.0 cm. An erythematous border could not be discerned. The mucous membrane, palms, and soles and nails were normal. The results of blood and stool examination including liver function tests were normal. The hemoglobin was 13 gm/dl. The skin biopsy showed focal vacuolar alteration of basal layer with mild to moderate infiltrate of lymphocytes and histiocytes intermixed with melanophages in the dermis, which was characteristic of ashy dermatosis and also lichen planus pigmentosus and other conditions with lichenoid histopathology [Figure 2]. A diagnosis of ashy dermatosis was made on the basis of the clinical and histological findings. The patient was advised photoprotection with a broad-spectrum sunscreen as well as tablet dapsone 25 mg daily after performing G-6-P-D estimation and liver function tests. A mild subjective decrease in the pigmentation was seen after 4 weeks of treatment. However, then the patient was lost to follow up.

DISCUSSION

Ashy dermatosis or erythema dyschromicum perstans (EDP) was first described by Oswaldo Ramirez in 1957. His patients were from Central and South America.[1] The term 'erythema dyschromicum perstans' was suggested by Dr. Sulzberger while examining Convit's patients with similar symptoms to those described by Ramirez, but, in addition had active lesions with raised erythematous margins.[2] Many authors consider ashy dermatosis and erythema dyschromicum perstans as a single entity, and the terms have been used interchangeably. Ashy dermatosis or EDP is an uncommon

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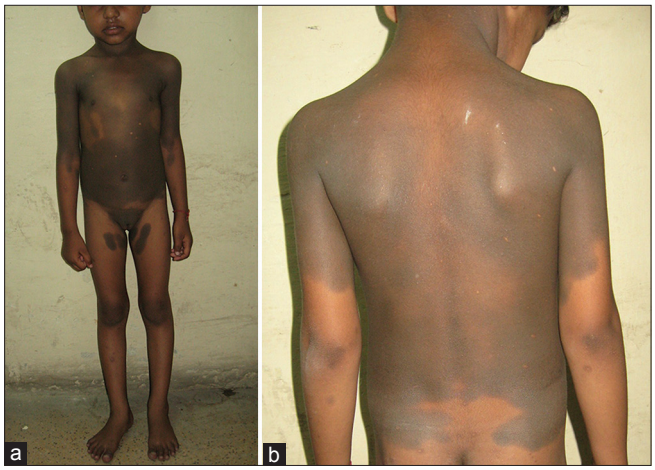

Figure 1: (a) Ashy gray-colored, confluent, symmetrical macules with polycyclic margins. (b) Lesions getting confluent with some areas being spared

entity, but appears to be more common in Latin America and Asia. Of the cases reported so far in the English language literature, it appears to be rare, and a total of 39 children have been described.[2] Unlike adult patients, who are mostly of Hispanic, Indian and South Asian origin, children with EDP are usually Caucasian.[3] In prepubertal children with EDP, there is absence of consistent trigger factors and an eventual improvement or resolution of the lesions in 50% cases.[2] In the Indian subcontinent, we have a similar simulating condition known as "lichen planus pigmentosus," which presents as hyperpigmented macules over the flexures and photoexposed areas.[4] The present case is reported because of the rarity of ashy dermatosis condition in childhood, especially in Asian population and also to stress upon the subtle differences between ashy dermatosis and lichen planus pigmentosus.

The etiology of ashy dermatosis or EDP is unknown. However, a number of etiological factors like ingestion of ammonium nitrite, nematodes infestation, radiographic contrast media, cobalt allergy, and chlorothalonil exposure among banana farm workers have been implicated.[5-8] None of these could be elicited in this patient. Women and darker-skinned individuals are more often affected.

Ashy dermatosis has been reported rarely in children. In the prepubertal patients, 52% cases of ashy dermatosis were Caucasians, 36% Hispanics, 4% African American, 4% Asian, and 4% unspecified.[3] To the best of our knowledge, only a single case report has been published of a case of ashy dermatosis in an Indian child.[9]

Onset is usually between the first to third decades of life. It starts abruptly or slowly as an ashy gray asymptomatic symmetric macular lesion of approximately 0.5 cm-2 cm over the trunk and spreads centrifugally to face and extremities. Early lesions have elevated erythematous active borders

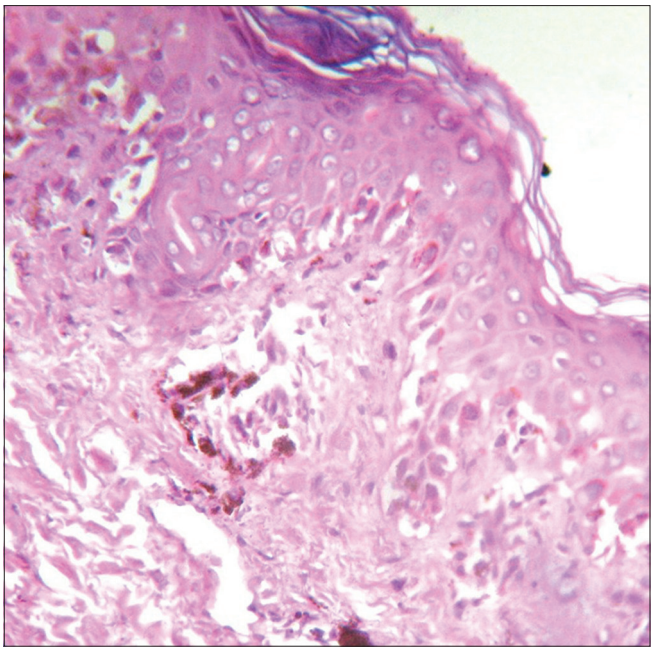

Figure 2: Vacuolar alteration of basal layer with mild to moderate infiltrate of lymphocyte to histiocyte intermixed with melanophages. (H and E, ×100)

about 1-2 mm. However, this border is often not present and eventually disappears after several months. Also, the erythematous border may not be perceptible in such dark-skinned patients. Lesions are oval, polycyclic, or irregular in shape. The palms, soles, scalp, nails, and mucous membranes are typically spared. Histology shows lichenoid dermatitis.[9,10] Direct immunofluorescence microscopy studies have demonstrated colloid staining of IgM and C4, and presence of fibrinogen at the dermoepidermal junction.[11]

Ashy dermatosis or EDP has a slow onset and is unlikely to resolve spontaneously. But, in children, it may be more likely to resolve within 2-3 years.[3]

Bhutani *et al*. described 40 Indian patients with similar symptoms to those reported by Ramirez, except that about one-third had associated lichen planus clinically and histologically.[4] This entity was called lichen planus pigmentosus, and it is considered a macular variant of lichen planus. Vega *et al*. reported in 1992 that LPP and ashy dermatosis are distinct entities and presented clinical differences between the two.[12] Some argue that ashy dermatosis may be a variant of LP. This was based on the clinical observation that EDP may accompany, precede, or follow lesions of LP and from the similarities shared on histology and immunofluorescence. The two disorders may be histologically indistinguishable.[4,13] LPP is differentiated by the distribution of lesions in photoexposed and flexures, presence of lesions of LP subsequently or simultaneously and the associated pruritus. On the other hand, EDP does not have any predilection for photoexposed areas. It is usually

not pruritic. LPP presents with patterns of pigmentation, which are diffuse (77.4%), reticular (9.7%), blotchy (7.3%), and perifollicular (5.6%) in nature.[13] However, in EDP, the lesion starts as a symmetrical ashy gray macules over the trunk that spreads in a centrifugal manner to involve the extremities with polycyclic margins. There can be residual hypopigmented halo in cases of EDP. LPP is a chronic condition with relapses and remissions, whereas EDP has a chronic and insidious course. Erythema is not easily perceptible in South Asian skin as in this patient, and hence the border may not have been noticed.

The therapeutic options for ashy dermatosis are clofazimine, dapsone, chemical peels, antibiotics, corticosteroids, vitamins, tetracyclines, anti-histamines, griseofulvin, isoniazid, chloroquine, and psychotherapy. However, none produces satisfactory results. In our setup, dapsone is a time-tested drug and is easily tolerated by the patients, hence it was given in this patient. We conclude by stating that EDP and LPP should be considered as distinct entities. Also, unlike adults, ashy dermatosis in children can have an eventual improvement or spontaneous resolution as was seen in a study in prepubertal children.[2,3]

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