Partial unilateral lentiginosis with Lisch nodules: A forme fruste of segmental neurofibromatosis?

Sir,

Partial unilateral lentiginosis is an unusual pigmentary disorder characterized by multiple lentigines in a unilateral distribution. The lesions often have a segmental pattern with a sharp demarcation at the midline. Histologically they are characterized by prominent rete ridges of the epidermis and an increase in the melanin content of the basal layer.\textsuperscript{1}

A 21-year-old female presented with multiple hyperpigmented macules on the left side of the face of 17 years’ duration. Hyperpigmented macules were noticed on the left side of the cheek at the age of 4 years, which gradually progressed to involve the left half of the face, neck, chest and upper back. No history of photosensitivity was present. There was no history of epilepsy, mental retardation or a family history of similar illness. The patient gave a history of easy fatiguability since childhood.

On examination, the patient was pale. Systemic examination revealed no abnormality. Cutaneous examination showed multiple, discrete, hyperpigmented macules of sizes varying from 2-5 mm on normal skin on the left half of the face, neck, upper chest with clear demarcation in the midline (Figure 1). There was no evidence of cafe-au-lait macules, axillary freckling or neurofibromas. The left eye showed hyperpigmented spots on the sclera. On ophthalmoscopic examination, both eyes showed Lisch nodules. Wood's lamp examination of the skin lesions showed accentuation of the pigmentation with no background hyperpigmentation.

On investigation, the patient had hypochromic microcytic anemia with hemoglobin of 8.5 g%. An electrocardiogram, echocardiography, and ultrasonography of the abdomen were within normal limits. A skin biopsy showed aggregates of melanocytes with increase in melanin in the basal layer and elongation of rete ridges suggestive of lentigines (Figure 2). Melanin bleach was positive.

The lesions of partial unilateral lentiginosis first appear during childhood. They can appear anywhere on the body but the upper extremities are more affected than the lower ones.\textsuperscript{1} The condition results from mutation during embryonic development probably confined to neural crest melanoblasts.\textsuperscript{2} It can be associated
with various central nervous system abnormalities including mental retardation,3 neurofibromatosis,4 cerebrovascular abnormalities with focal epilepsy,2 and probably with iron deficiency anemia and euthyroid goitre.1

The coexistence of neurofibromatosis with partial unilateral lentigines raises the possibility that partial unilateral lentigines could be a variant or forme fruste of segmental neurofibromatosis.2 Our patient had no cutaneous lesions of neurofibromatosis but had bilateral Lisch nodules in the eyes. Since Lisch nodules are characteristic of neurofibromatosis, we conclude that the partial unilateral lentiginosis seen in our patient is a forme fruste of neurofibromatosis.

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REFERENCES

Peeling skin syndrome

Sir,

Peeling skin syndrome (PSS) is a rare disorder characterized by continual shedding of the entire stratum corneum. It starts either at birth or later in childhood.1 It is inherited in an autosomal recessive pattern.2 The skin involvement is usually generalized, but in some patients sparing of the face, palms and soles has been reported.3 Here we report a case of PSS in a girl who had sparing of palms and soles and whose disease showed summer exacerbation.

A 5-year-old girl presented with a history of peeling of skin from all over the body in patches since 1 year of age. She was born of a non-consanguineous marriage and there was no history of a similar disorder in any of her family members. Her problem of peeling of the skin remained all around the year but worsened in the summer. The lesions were mildly itchy. On examination she had patches of peeling skin all over the body along with a few lesions on her face (Figure 1). Rubbing the normal skin revealed the peeling but there was no oozing from the rubbed area. Some hyperpigmented patches remained at the sites of old lesions. The palms, soles and mucous membranes were uninvolved. General and systemic examination revealed no abnormality. Routine hemogram and urinalysis were normal. Histopathological examination of the skin showed separation of the stratum corneum away from the stratum granulosum, mild acanthosis and a normal dermis (Figure 2). The patient was treated with emollients which gave her symptomatic relief to some extent, but on follow up she had recurrence of the lesions.

PSS is variously known as keratolysis exfoliativa, congenital deciduous skin, and familial continual skin peeling.4 Troupe has pointed out that there are two types of peeling skin syndrome. The first was first