DERMATO-VENEREOLOGICAL QUIZ

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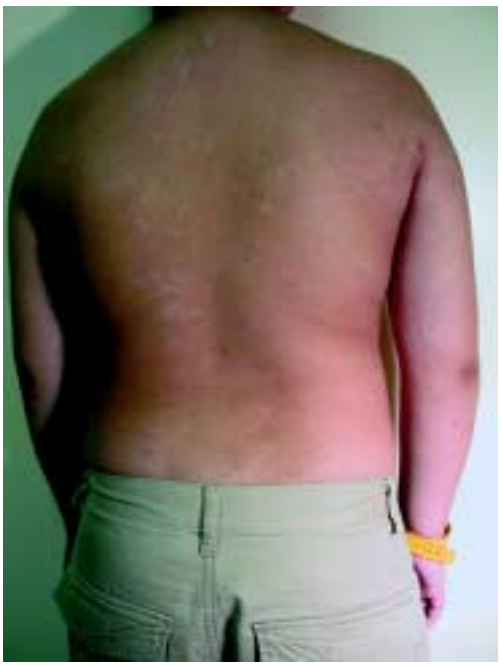

Figure 1

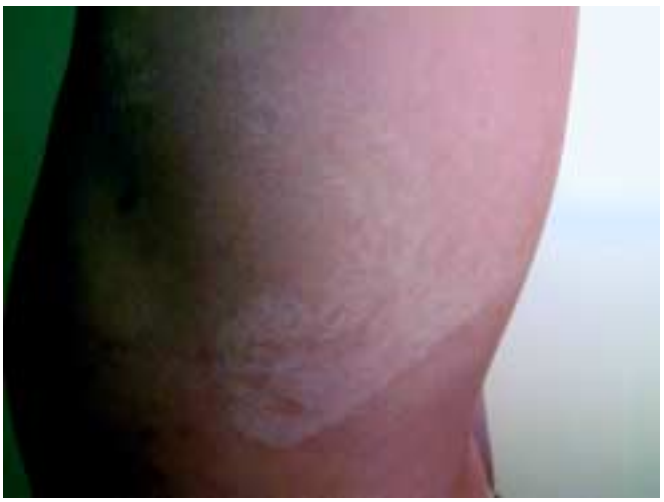

Figure 2

Question 1

This 9-year-old Chinese boy was noted to have streaky hypopigmentation over his body and limbs, both anteriorly and posteriorly, since infancy. There was no history of itchy rash, blistering, abnormal growth or hyperpigmentation over the site of involvement. He enjoys good health and is excellent at school.

1. What is the likely diagnosis?
2. What will one see on histological and electron microscopic examination?
3. What are the typical clinical presentations of this condition?

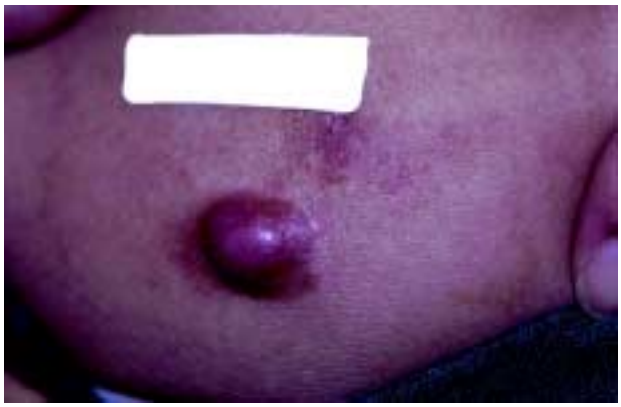

Figure 3

Question 2

This 39-year-old lady noted a slowly enlarging asymptomatic nodule over her abdomen for the past two years. Examination revealed a hard, raised dermal nodule with surrounding induration. There was another similar but smaller nodule situated superiorly.

1. What are the differential diagnoses?
2. What are the typical histological findings?
3. What is the treatment for this lady?

(answers on page 104)

Answers to Dermato-venereological Quiz on page 97

Answer (Question 1)

1. The pictures show whorled hypopigmented streaks over the body and limbs bilaterally. Together with the history, the most likely diagnosis is hypomelanosis of Ito.

2. In the hypopigmented areas, Fontana-Masson stain shows a decrease in, or complete absence of, melanin granules in the basal cell layer. With the dopa reaction, the hypopigmented areas are seen to contain fewer and smaller melanocytes than normal, with sparse, short dendrites. There is no pigmentary incontinence, in contrast to incontinenti pigmenti. Electron microscopy shows a significant decrease in the number of melanosomes within the melanocytes and keratinocytes. In addition, some melanocytes may show degenerative changes and an absence of melanosomes.

3. Hypomelanosis may be noted at birth, but it may not begin until childhood. There may be partial repigmentation in adulthood. The characteristic skin sign is a whorled pattern of hypopigmentation of variable extent, which may be unilateral or bilateral, along the Blaschko's line. Congenital abnormalities are found in about 50% of the cases. The most common associations are mental retardation and seizure disorders, but there may also be abnormalities of the eyes, hair, teeth or the musculoskeletal system.

Answer (Question 2)

1. The differential diagnoses include dermatofibrosarcoma protuberans (DFSP), dermatofibroma and keloid.

2. DFSP is composed of densely packed, monomorphous, plump spindle cells arranged in a storiform pattern in the central areas of tumour nodules, whereas at the periphery there is diffuse infiltration of the dermal stroma, frequently extending into the subcutis. In contrast to dermatofibroma, the epidermis overlying DFSP is usually attenuated rather than hyperplastic. Immunostaining for CD34 is usually positive in DFSP and negative in dermatofibroma.

3. This lady has DFSP. The lesion should be excised completely, with at least a 5 cm margin. Local recurrence is not uncommon, as it is difficult to judge the lateral extent of the tumour.