CASE REPORTS

OSTEOMA CUTIS

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A case of primary osteoma cutis seen in a 15-month-old female child is reported.

Key words : Osteoma cutis.

Cutaneous ossification or bone formation in the skin is a very rare condition. It is either primary or secondary. Secondary cutaneous ossification occurs through metaplasia in a pre-existing lesion which may be either a tumor, a scar or an old inflammatory process. Primary cutaneous ossification is still rarer. Lever[1] has defined it as cutaneous ossification without any preceding lesion. He has classified it into two categories : (a) Albright's hereditary osteodystrophy, and (b) osteoma cutis. According to Brooke and Valman,[2] Albright's hereditary osteodystrophy, first described in 1952, consists of multiple areas of subcutaneous or intracutaneous ossification. These are present at birth or may appear later in life. There is no definite area of predilection. The size may vary from a minute area to as large an area as 5 cm in diameter. These may ulcerate through the skin. In some cases, bone formation is seen in the fascial planes also, in addition to ossification in the skin. Albright's hereditary osteodystrophy includes the syndromes of pseudohypoparathyroidism and pseudopseudohypoparathyroidism. The patients are usually short statured, have a round facies and also have multiple skeletal abnormalities. Albright's dimpling sign is positive. Additional features may include calcification of the basal ganglia and mental retardation. The mode of inheritance seems to be dominant and possibly it is X-linked.

The second variety of primary cutaneous ossification is osteoma cutis. In this case, there is no evidence of Albright's hereditary osteodystrophy in either the patient or the family. Apparently four groups with osteomas have been described by Peterson and Mandel.[3] The osteomas are limited in extent except in the first group. The four groups consist of : (a) patients with widespread osteomas since birth or early life, (b) patients with a single large plaque-like osteoma present since birth either in the skin or scalp, (c) patients with a single small osteoma arising in later life in various locations, (d) patients with multiple miliary osteomas of face. This variety is seen exclusively in the females. In some cases, the osteomas do not appear until late in life. According to Basler et al,[4] association with acne seems to be purely coincidental.

We are reporting a case of primary osteoma cutis.

Case Report

A 15-month-old female child was seen with

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yellowish, papulo-nodular lesions extending from the right knee to the right ankle (Fig. 1).

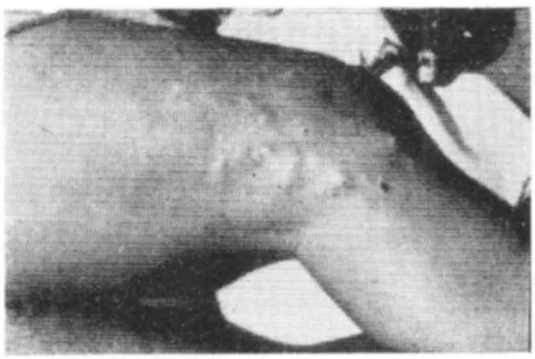

Fig. 1. Papulo-nodular lesions on the right thigh, knee and leg.

The lesions were first noticed when the child was 2-week-old. In addition, there were firm plaques on the lateral aspects of the knees, the thighs and the legs. There was no ulceration. The joints were not involved. The gait was normal. The child is a product of non-consanguinous marriage. The mental milestones were normal. There were no other systemic abnormal findings.

A biopsy from the lesion on the right leg showed well-formed laminated bone with Haversian canal in the mid and lower dermis. At the periphery of the bone there were two areas of calcification

An X-ray picture of the right lower extremity showed diffuse radio-opaque shadows of cutaneous ossification Biochemical investigations on blood revealed no abnormality. Serum calcium was 8.8 mg%, serum alkaline phosphatase was 4 KA units, and serum acid phosphatase was 2 KA units

Comments

As the child's milestones were normal, there were no other skeletal abnormalities, the Albright's dimpling sign was negative and neither the patient nor her relatives showed any clinical evidence of Albright's hereditary osteodystrophy, we feel that this is a case of primary osteoma cutis.

References

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