A Case of Primary Osteoma Cutis

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Primary osteoma cutis in an otherwise healthy 7-month-old male infant was presented as several hard plaques on the trunk and the extremities. Histologically, lesions revealed bony spicules with numerous osteocytes. The history and the result of a physical examination failed to reveal any skin lesion prior to the development of the lesions. There was no evidence of Albright's hereditary osteodystrophy in either this patient or his family.
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Key Words : Primary osteoma cutis, Albright's hereditary osteodystrophy

Osteoma cutis, which is characterized by dermal or subcutaneous bone formation, is a rare cutaneous disorder. Pathogenetically, cutaneous ossification may be classified as primary or secondary.This classification is based on the presence or absence of a preceding cutaneous lesion[1]. Primary cutaneous ossification is much less common than the secondary form and arises idiopathically, typically showing a benign true new bone formation composed of mature, lamellar and/or osteonic bone tissue within the deeper dermis or in the subcutaneous tissue[2]. Mature, true bone formation has not been described in secondary ossification. Therefore Cottoni et al[3]. proposed that the term "secondary mineralization" may be more suitable than secondary ossification. The term "osteoma cutis" is applied to cases of primary cutaneous ossification in which there is no evidence of Albright's hereditary osteodystrophy in either the patient or his family[4,5].

Two cases of osteoma cutis have been reported in Korea[6,7]. We now describe the clinical and histological features of an additional case of primary osteoma cutis in an otherwise healthy 7-month-old male infant.

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REPORT OF A CASE

A seven-month-old male infant visited our clinic. His chief complaint was a stony hard cutaneous lesion on his left shin. His mother had noted the mass first when the patient was four months old. Since then it had become more prominent and increased in size. There was neither the history of trauma nor of previous skin disorders. No family history of similar skin lesions or other hereditary conditions was found. The history of his mother's pregnancy was unremarkable.

During a physical examination, relatively well defined, 6 × 3.5cm sized, stony-hard, slightly reddish plaque, adherent to overlying skin, was seen on his left shin area(Fig. 1).

A examination by a pediatrician had showed no abnormality. His growth and development were normal.

The biopsy specimen was obtained under local anesthesia from the lesion of his left shin. Routine H and E staining revealed calcification and ossfication with many osteocytes lying within the lacunae and a few osteoblasts in the deep dermis, but no osteoclast. The surrounding tissue was entirely normal and no inflammatory cells were present(Fig. 2, 3).

Results of the laboratory investigations including serum levels of calcium, phosphorus, alkaline phosphatase, and parathyroid hormone were ei-

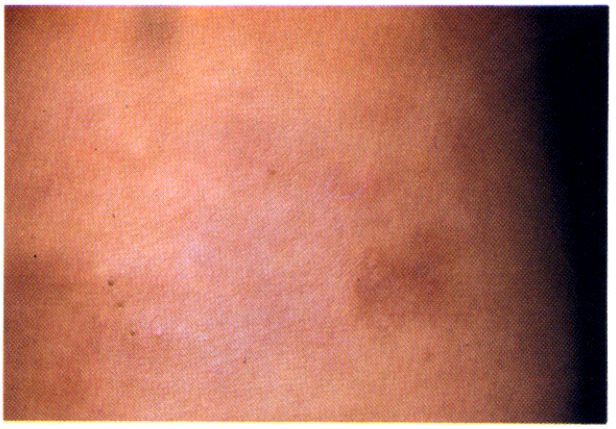

Fig. 1. 6 × 3.5 cm sized slightly erythematous, irregularly shaped, hard plaque on left lower leg.

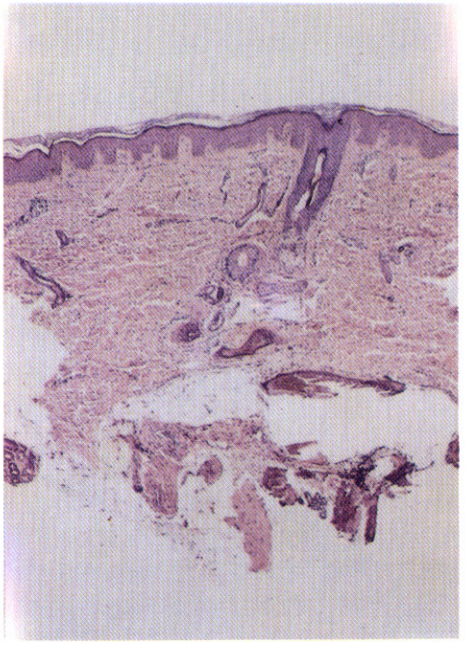

Fig. 2. A low power view of a biopsy specimen, showing osteoid bone formation and calcification in the deep dermis and subcutaneous tissue(H & E, 40).

ther negative or within normal limits except for a mild elevation of SGOT/SGPT(70/57).

About 3 weeks after the initial examination, several new lesions developed on his lower back and right forearm. An examination of his skin revealed pea sized subcutaneous nodules which showed the same features of the previous left shin lesion. The patient was referred to a plastic surgeon for surgical excision.

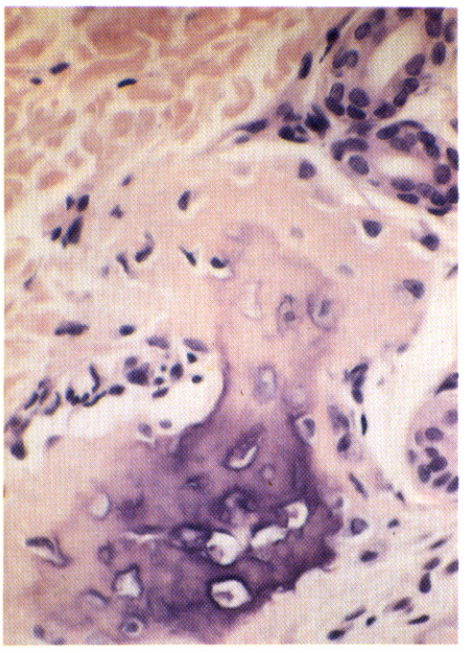

Fig. 3. Bony spicules containing numerous osteocytes(H & E, 400).

DISCUSSION

Bone formation within the skin may be primary or secondary. Primary cutaneous ossification can occur in Albright's hereditary osteodystrophy (AHO) and as primary osteoma cutis. Secondary, metaplastic ossification occurs in association with a wide variety of conditions, including cutaneous tumors, nevi, scars or during an inflammatory process[8,9,10]. Localized lesions of primary cutaneous ossification are often referred to as "osteoma cutis". Since the first description of osteoma cutis by Wilkins[11] in 1858, there have been many additional cases reported in the literature, but only a few have been primary osteomas. The term "osteoma cutis" is restricted to cases of primary cutaneous ossification in which there is no evidence of AHO in either the patient or his family[4,5]. Roth et al[10]. preferred the term "cutaneous ossification" to bypass the difficulty of identifying a pre-existing lesion by histological means.

The etiology and pathogenesis of this disease still remain unclear. It has been suggested that the osteoblasts and osteocytes in primary osteoma cutis originate from mesenchymal cells[12]. O'Donnel and Geller[13] proposed that primary osteoma

cutis is a membraneous osseous hamartoma which arises de novo, not requiring local tissue or systemic chemical abnormalities for its formation. Recently Oikarinen et al.[14] suggest that fibroblasts have the ability to differentiate into osteoblastic cells, which have some properties of osteoblasts such as high alkaline phosphatase activity and a high expression of osteonectin.

Primary osteoma cutis is a rare skin tumor, with no tendency for invasive growth or metastasis. These benign tumors result from a new growth of bony tissue within normal dermis. The diagnosis of primary osteoma cutis may be suspected clinically by the stony consistency of the lesions but it can be made only by histologic examination. The lesions appear as hard, round to irregular, sharply defined tumors of varying size. The color ranges from flesh-colored to purple or brown, and the lesions are usually situated deep in the dermis or subcutaneous tissue[1]. Microscopic examination reveals spicules of bone of varying sizes in the dermis and subcutaqneous tissue. The bone contains numerous osteocytes as well as cement lines. Osteoblasts may be found at the edge of the bone spicules. Haversian canals are also often present[4].

In our case, there was no evidence of any preceding inflammation or of cutaneous disorder such as trauma, injection, tumor, or nevus, although it was difficult to exclude the possibility of previous unrecognized trauma with certainty. Furthermore, the calcium/phosphorus balance and the serum level of the parathyroid hormone were within normal limits, excluding Albright's syndrome. AHO includes both pseudohypoparathyroidism and pseudopseudohypoparathyroidism. Patients with AHO are short of stature, have round face and soft tissue calcification. The associated skeletal abnormalities in AHO include curvature of the radius and exostosis, as well as shortened metacarpal and metatarsal bones[15,16]. The case presented in this paper had no clinical evidence of AHO and no family hitory of AHO. Histopathologic examination was also compatible with histologic findings of primary osteoma cutis. Therefore the case was considered to be a primary osteoma cutis.

Mineralized areas of primary osteoma cutis may be divided into macrocalcification and microcalcification[3]. Macrocalcification is completely calcified plaque composed of lamellar bone and microcalcification is dispersed around the calcified plaque composed of osteoid tissue. During light microscopic examination, it was difficult to decide on the type of mineralization in this case because there were calcified plaques without any lamellar bone structure. The absence of lamellar bone structure may be due to the immaturity of the cutaneous ossification.

Lever and Schaumberg Lever[4] divided primary ossification into four groups. The four groups are (1)patients with widespread osteomas since birth or early life but without any evidence of AHO; (2)patients with a single, large, plaquelike osteoma present since birth either in the skin of the scalp or in the skin or subcutaneous tissue of an extremity; (3)patients with a single small osteoma arising in later life in various locatons and in some instances showing transepidermal elimination of bony fragments; (4)patients with multiple miliary osteoma of the face. Some feel the miliary osteomas are metastatic because they may be occurring in acne scars[17]. Our case may be considered to be a widespread osteoma because several new lesions developed on the trunk and the upper extremities.

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