

Adult onset plate-like osteoma cutis

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Abstract:

Cutaneous ossification refers to the rare occurrence of bone in the skin. It may be primary, occurring in normal skin, or secondary, occurring in disrupted skin tissue. Primary osteoma cutis may represent an independent phenomenon or may be part of a syndrome ⁽¹⁾. A 50-year-old woman had a stony-hard plaque on her scalp 25 years ago. The histological features were those of osteoma cutis without underlying skin pathology. She had no predisposing conditions such as trauma or relevant dermatologic diseases. The patient described herein had a unique presentation of adult onset primary plate-like osteoma of the scalp which is a rare variant of osteoma cutis.

Introduction:

Cutaneous bone formation is an unusual event that is traditionally divided into primary and secondary types (Table 1) ⁽²⁾. Secondary osteomas account for 85% of cutaneous ossifications and develop usually within pre-existing inflammatory, neoplastic and non-neoplastic skin lesions. Primary osteoma cutis accounts for 15% of cutaneous ossifications and includes cases associated with Albright's hereditary osteodystrophy (AHO) and those occurring as isolated defects without such association ⁽³⁾.

The term osteoma cutis is usually applied to cases of primary cutaneous ossification in which there is no evidence of AHO ⁽⁴⁾. Osteoma cutis is divided into four clinical types: isolated osteoma, widespread osteomas, multiple miliary osteomas of the face, and plate-like cutaneous osteoma which had previously described as dysplastic cutaneous osteomatosis and osteosis cutis ^(5,6,7). Histopathologically, osteoma cutis shows osseous tissue with trabeculae enclosing fat, and occasionally bone marrow cells in the dermis and subcutaneous tissue ^(8,9,10).

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We report a case with adult-onset plate-like osteoma cutis that usually is described in neonates and considered to be congenital.

Case report:

A 50-year-old woman presented with a slowly growing, asymptomatic skin lesion on the right temporal area of the scalp of 25 years duration. On physical examination, the lesion was solitary well-circumscribed, stony-hard irregular plaque. It was reddish yellow in color and of about 7 cm in width (Fig. 1 A&B). There was no history of trauma or any previous cutaneous lesion on the scalp. She did not have phenotypic features of AHO and no family history of similar skin lesion.

Various investigations including complete blood counts (CBC), serum calcium and phosphorus, serum electrolytes, as well as liver and renal function tests were all within normal limits. Her skull X-ray revealed a bone thickening along the inner cortex of the right parietal bone (Fig. 2).

Skin biopsy specimen from the lesion showed irregular trabeculae of lamellar bone within the dermis extending to the subcutaneous fat (Fig. 3). The trabeculae were lined by osteoblasts (Fig. 4). The overlying epidermis appeared normal. No underlying cutaneous pathology was observed.

Based on the medical history, clinical evaluation, serum chemistry profile and histopathologic findings, the diagnosis of adult onset plate-like osteoma cutis was confirmed.

Discussion:

Cutaneous ossification is classified as primary when it arises *de novo* or secondary when it develops within a pre-existing skin lesion ⁽¹¹⁾. The causes of cutaneous ossification are summarized in Table 1.

In our patient, there was no evidence of a secondary process within the biopsy specimen, which ruled out the possibility of secondary cutaneous ossification, as occurs in tumors, nevi, or other inflammatory conditions. Furthermore, clinical signs and serological tests that suggest a collagen vascular disorder e.g. systemic sclerosis were absent. By a combination of history, clinical features, laboratory data, and histopathological findings, the condition is categorized as primary osteoma cutis. Primary cutaneous osteomas are extremely rare. In a retrospective analysis of 20,000 consecutive biopsies of the skin, Burgdorf and Nasemann ⁽¹²⁾ found only 10 primary cutaneous osteomas ⁽¹³⁾.



Fig. 1:
A) A reddish yellow plaque on the right temporal region of the scalp.
B) The plaque is well defined and has irregular surface.

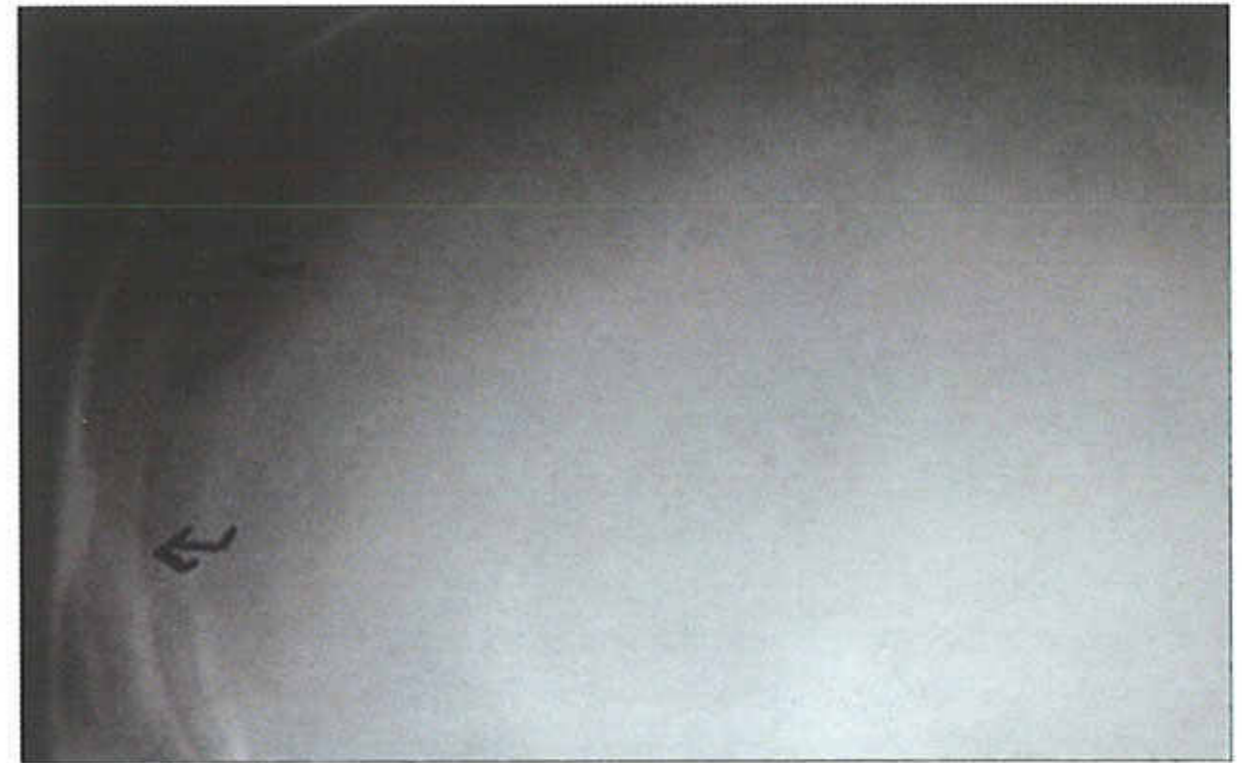


Fig. 2. Skull X-ray showing thickening along the inner cortex of the right temporal bone.

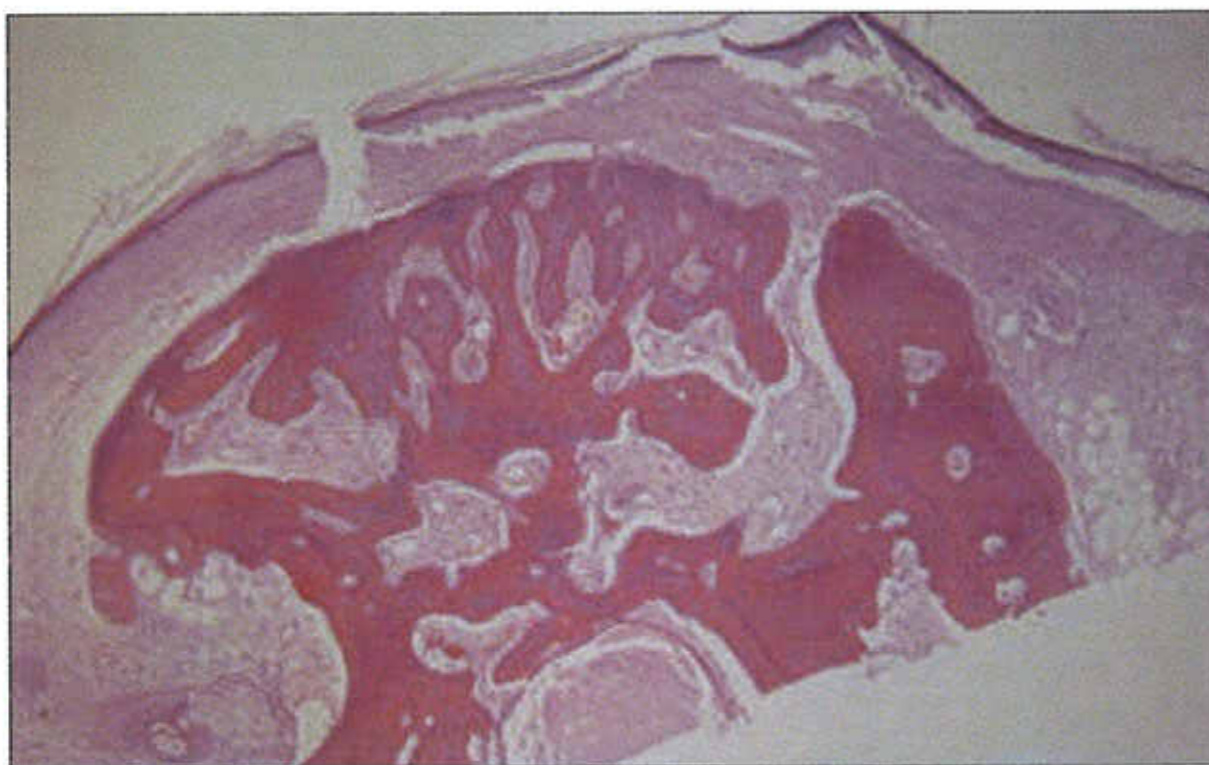


Fig. 3. Irregular trabeculae of lamellar bone occupying the dermis and extending deep into subcutaneous fat.

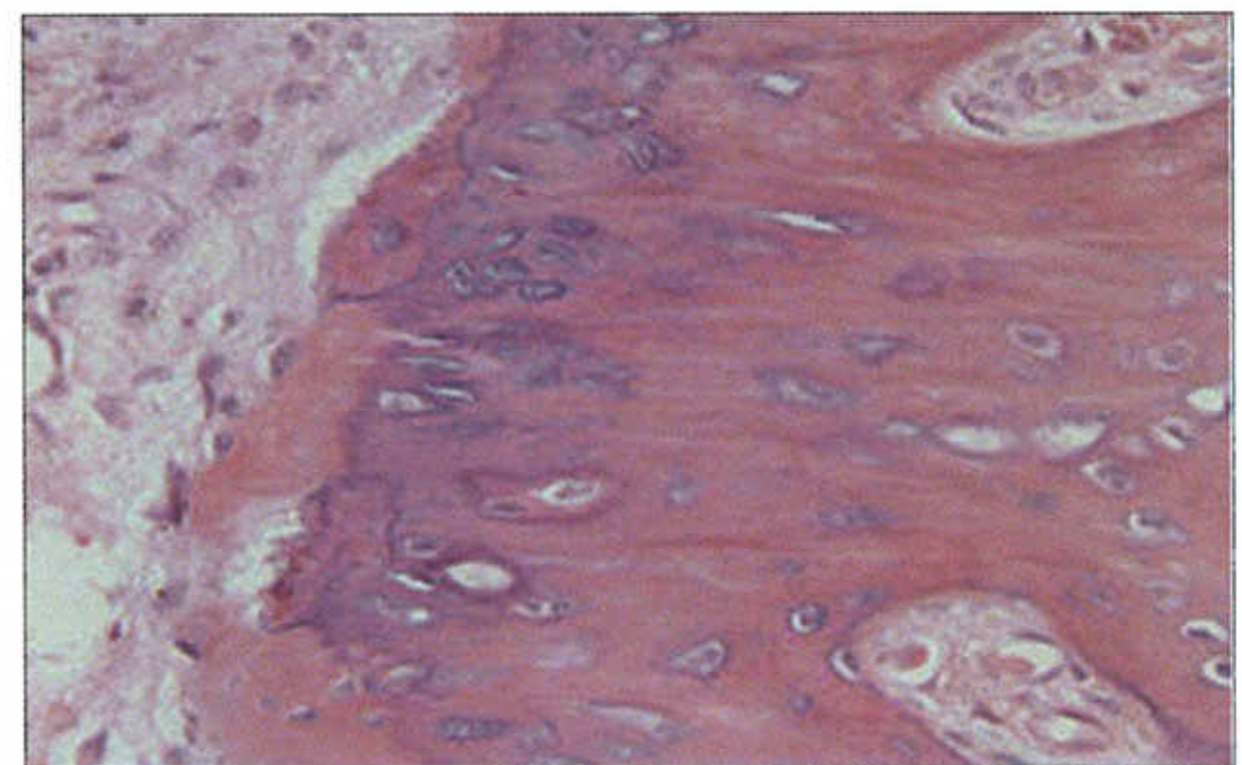


Fig. 4: Bone trabeculae lined by osteoblasts.

Primary cutaneous osteomas are reported to be associated with Albright's hereditary osteodystrophy (AHO). AHO is a syndrome with a heterogeneous mode of inheritance and characterized by endocrine, skeletal, neurologic, and cutaneous disturbances⁽¹⁴⁾. Patients with AHO typically have a deficient end organ response to parathyroid hormone with hypocalcaemia, hyperphosphatemia and elevated serum parathyroid hormone level⁽¹⁵⁾. AHO phenotypic features include round facies, short stature, obesity, numerous skeletal abnormalities⁽⁵⁾. So, AHO was excluded, in our case, by physical and laboratory evaluation.

Different forms of osteoma cutis are detected by their location, appearance and number. However, not all osteoma cutis cases can be clearly categorized. Osteoma cutis is divided into four clinical patterns: (a) Widespread osteomas which present in the neonatal period as multiple generalized subcutaneous osteomas. (b) Multiple

miliary osteomas of the face which usually occur in women with or without history of acne. (c) Single osteoma arising later in life which presents as a single isolated nodule at any cutaneous site. (d) Congenital plate-like osteoma which occurs in neonatal period or during first year of life. It presents morphologically as a large plaque on the scalp or the extremities^(5, 16). Some acquired cases have been described secondary to previous inflammatory skin disorders like morphea which was pathologically absent in this case^(5, 17). The onset of the lesion, in our patient, is unusual as it was noticed at the age of 25 which excludes congenital primary cutaneous ossification. Thus we report a case of adult onset plate-like osteoma cutis.

Mechanism of cutaneous ossification is unclear. However, it has been speculated that the bone deposition is either due to the ability of either resident fibroblasts or nests of pluripotent mesenchymal cells to dif-

ferentiate into osteoblasts¹². The alkaline phosphatase activity as well as the expression of calcium binding glycoprotein, "osteonectin" are observed to be increased in osteoma cutis lesions^(18,19).

The prognosis of plate-like osteoma cutis is generally considered good. Malignant transformation of cutaneous osteomas has not been described as yet. The treat-

ment of these cases is not settled yet. However, surgical interference is the treatment of choice in most cases. Several authors have tried medical treatment with diphosphonate (etidronate disodium) therapy without significant results^(1,3,16,20). Regarding our case, the recommended surgical management was refused by the patient and her family.

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Table 1: Causes of cutaneous ossification⁽⁵⁾

<p>1. Primary</p> <ul style="list-style-type: none"> a) Albright's hereditary osteodystrophy b) Primary osteoma cutis (No association with Albright's hereditary osteodystrophy) <ul style="list-style-type: none"> 1) Isolated osteoma 2) Widespread osteoma 3) Multiple miliary osteomas of the face 4) Plate-like osteoma <p>2. Secondary</p> <ul style="list-style-type: none"> a) Inflammatory skin disease <ul style="list-style-type: none"> 1) Progressive systemic sclerosis and CREST syndrome 2) Dermatomyositis 3) Morphea b) Tumor <ul style="list-style-type: none"> Basal cell carcinoma, pilomatrichoma, etc. c) Trauma and scars
