Case Report

Two cases of primary osteoma cutis occurring in children

兩例兒童的原發性皮膚骨瘤

CH Song 宋翠豪, LQ Zheng 鄭力強, ZG Zhao 趙梓綱, W Ba 巴偉, L Wang 王嵐, CX Li 李承新

We report two cases of primary osteoma cutis in children. The lesions were all located on the legs, presenting as hard nodules covered by normal or mildly erythematous skin. The histopathology showed well-formed spicules of bone in the dermis. They were classified as primary osteoma cutis because there were no pre-existing lesions.

We報告了兩例兒童的原發性皮膚骨瘤。病變均位於腿部，表現為皮下多發硬結節。組織病理學顯示真皮內成熟的骨組織。本例皮損發生前局部無其他皮膚損害，原發性皮膚骨瘤診斷明確。

Keywords: Histopathology, Neoplasm, Primary osteoma cutis

關鍵詞：組織病理學、腫瘤、原發性皮膚骨瘤

Introduction

Osteoma cutis is a rare benign dermatological disorder in which there is bone formation in the dermis or subcutaneous tissue.\(^1\) It can be divided into two types: primary and secondary. Secondary osteoma cutis, which is more common, develops within or adjacent to areas of inflammation, naevi or tumour, while primary osteoma cutis is not secondary to any other skin diseases.\(^2\) In a retrospective study of 425 cases of cutaneous ossification, only 14% of cases were primary.\(^3\) It is a rare condition and may be unfamiliar to the clinician. We report two cases of primary osteoma cutis (POC) in children and review the literature to gain some insight into this rare skin condition.

Case report

Case 1

A 9-year-old boy presented with multiple nodules on the right knee. The lesions had been present since he was three months old. His mother noted...
a small protuberance at first. There was some erythema initially which progressed to a skin colour with some atrophy. On physical examination, there were multiple hard nodules in the subcutaneous distributed in a 0.5 cm x 1 cm area. There was no adherence to surrounding tissues and no tenderness (Figure 1A). There was no family history of this condition or any history of drug or food hypersensitivity or left thigh injury and there were no developmental or intellectual abnormalities. The skin biopsy showed multiple ossification, osteoblast and calcification foci in the dermis (Figure 1B & 1C).

**Case 2**
A 4-year-old girl presented with a two-year history of a large, well-defined, mildly erythematous lesion on the left thigh. The lesion was hard and 10 cm in size, mobile, and was associated with spontaneous pain, tenderness (Figure 2A). There was no previous trauma and no other skin lesions were present at this site. The lesions developed in the absence of any known triggering factors. Skin biopsy study showed extensive formation of mature ectopic bone in the deep dermis with osteoblast and calcinosis (Figure 2B & 2C). These foci were circumscribed and heavily calcified.

**Discussion**
Osteoma cutis is rare benign dermatological condition caused by the deposition of bony nodules in the dermis. The disease can occur at any age, but most often affects children. It may be sporadic or multiple and lesions are often located on the scalp, forehead, cheek, and chin regions, but can also occur anywhere in the body. The lesions are often solid and round, with geographic borders and the overlying skin may be normal, erythematous, pigmented, atrophied or ulcerated. The diameter varies from 0.1 cm to 5.0 cm. The lesions are asymptomatic in most cases.

Histopathology is the gold standard for diagnosis. Typical histopathological findings include the presence of amorphous basophilic deposition in the subcutaneous fat and bone formation in the dermis. Osteoblast and osteoclast are seen within the lacunae. Sometimes there is bone lamellae with Haversian canals.

The pathogenesis of osteoma cutis is still unclear. Burgdorf and Nasemann have proposed two theories. The first suggests the anomalous migration of osteoblasts to the skin while the second theory involves the metaplasia of fibroblasts to osteoblasts. Some cases of POC are linked to GNAS Mutations. The GNAS1 gene encodes the $\alpha$-subunit of the stimulatory G protein of adenyl cyclase (Gs$\alpha$) and inactivating mutations of GNAS1 in patients lead to suboptimal activity of the Gs$\alpha$-activated pathways and result in heterotopic ossification of soft connective tissues.

There have been reports of Albright hereditary osteodystrophy (AHO) associated with POC, in which there was short stature, round face, obesity, brachydactyly and osteoma cutis. Mental retardation has been less frequently described in patients with AHO. In our two cases, there was no clinical evidence of association with these syndromes. However, AHO should be ruled out if there are multiple cutaneous ossifications in a young child.

The prognosis of osteoma cutis is good and is not associated with malignant transformation. Localised forms generally do not undergo significant growth. The treatment options osteoma cutis are varied and include topical retinoids or ablative lasers. Emel Bulbul Baskan reported the needle micro-incision-extirpation method, which is a simple, easy and inexpensive method that was first suggested by Thielen. Surgical treatment is still the most frequent reported treatment modality.
Figure 1. (A) Presence of skin coloured, hard, multiple nodules. (B) Skin biopsy showing the presence of bone formation in the dermis (HE 4x). (C) Osteoblast and calcification can be seen (HE 10x).

Figure 2. (A) Large, well-defined, mildly erythematous lesion on the left thigh. (B) Photomicrograph showing extensive formation of mature ectopic bone in the deep dermis with focal areas of adipose tissue between bony spicules (HE 4x). (C) Osteocytes can be found within the lacunae. At the periphery, there are compressed collagen fibres (HE 10x).
We report two cases of childhood POC. The lesions were all located on the legs, which is a relatively unusual site. The lesions were all with hard nodules, covered by normal or mildly erythematous skin. There was spontaneous pain in one case while the lesions were asymptomatic in the other. The histopathology showed well-formed spicules of bone in the dermis in both cases. They were classified as primary osteoma cutis because there were no pre-existing lesions.

In conclusion, POC is a rare condition of which dermatologists should be aware. We present two such cases to increase our understanding of this entity.

Declaration

Fund program: National Natural Science Foundation of China (81572680).

References