Case Report

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Pathology Section

Multiple Osteoma Cutis of the Wrist and Hand

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ABSTRACT

Osteoma cutis is a rare, benign condition characterised by dermal or subcutaneous bone formation that presents as stony hard nodules. The disease is classified into primary and secondary forms. Primary Osteoma Cutis (POC) is not associated with any predisposing conditions, whereas Secondary Osteoma Cutis (SOC) occurs as a result of metabolic conditions that lead to an increase in blood calcium levels. Here, author present a case of a 21-year-old girl having multiple tiny stony hard nodules on the right wrist and hand. Syndromic association was ruled out by obtaining a thorough clinical history, accompanied by physical examination and search for laboratory signs of pseudohypoparathyroidism. This ultimately led to a more accurate diagnosis of isolated primary osteoma cutis, a very rare condition reported in the literature.

Keywords: Cutaneous nodules, Heterotopic ossification, Multiple osteomas

CASE REPORT

A 21-year-old female presented to the Dermatology OPD with multiple (5-6) hard cutaneous nodules on the dorsum of her right hand. The lesions were first noticed by her when she was around 10-year-old. These nodules had started increasing in size since the last 2-3 years. Additionally, she developed dull pain in them since the last two years when the pressure was applied as while holding tailoring scissors. She was born healthy and there was no family history of similar lesions. The physical examination yielded a height of 139.7 cm (normal for an average Indian woman) and weight of 49.6 kg (normal for age). The cutaneous examination showed multiple, discrete, skin-coloured, hard, mildly tender papules, size ranging from 0.25 cm to 0.5 cm localised on the ulnar border of the wrist, dorsum of the right hand on the fingers and in the web spaces. No exudation of fluid or material was seen. On palpation, they were bony in consistency. The patient was also found to have a weak right limb Grade 2 and circumduction gait. She gave history of developing weakness of the right leg during infancy as told by her mother.

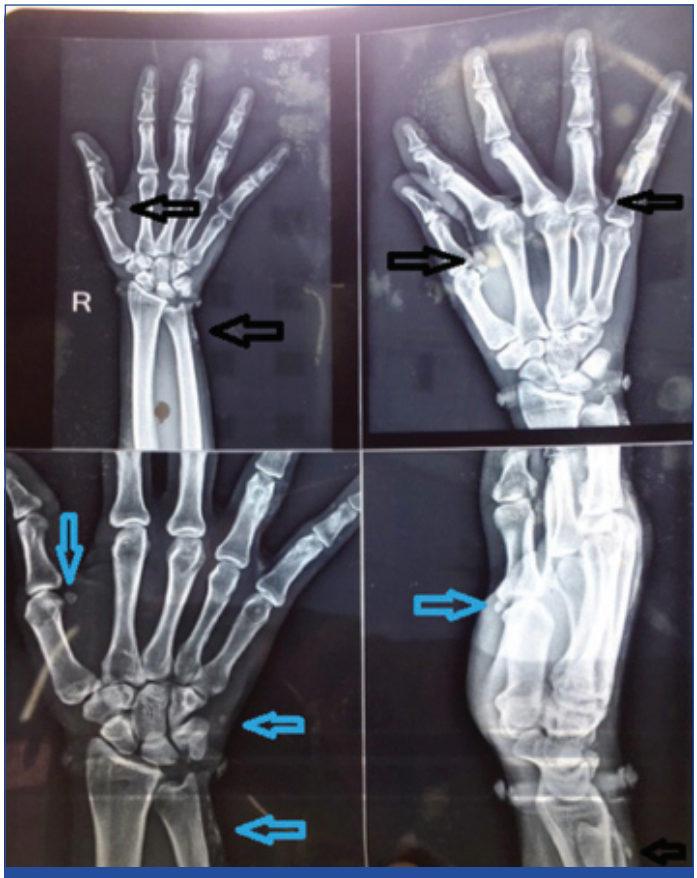
Radiographic examination of the right wrist and hand demonstrated well-defined bony nodules in the soft tissues of the right wrist and hand region [Table/Fig-1]. Radiographic examination of the limb did not reveal any evidence of ossification in the pelvis and limb bones. Blood investigations revealed a normal serum alkaline phosphatase level of 67 IU/L, a normal serum calcium level of 8.6 mg/dL, and blood phosphorus was also within normal limits at 8.6 mg/dL. Serum 25-hydroxy/vitamin D was decreased to 18.56 nmol/L. Serum parathormone levels were also found to be normal at 29 pg/mL.

The present patient had normal weight and height for age. Examination of the fingers and toes yielded no abnormality. Radiologically, absence of ossification in the deeper connective tissues helped to rule out progressive osseous heteroplasia. The limb shortening could be attributed to probable poliomyelitis in childhood. On these bases as well as based on the biochemical investigations, syndromic causes of POC were ruled out.

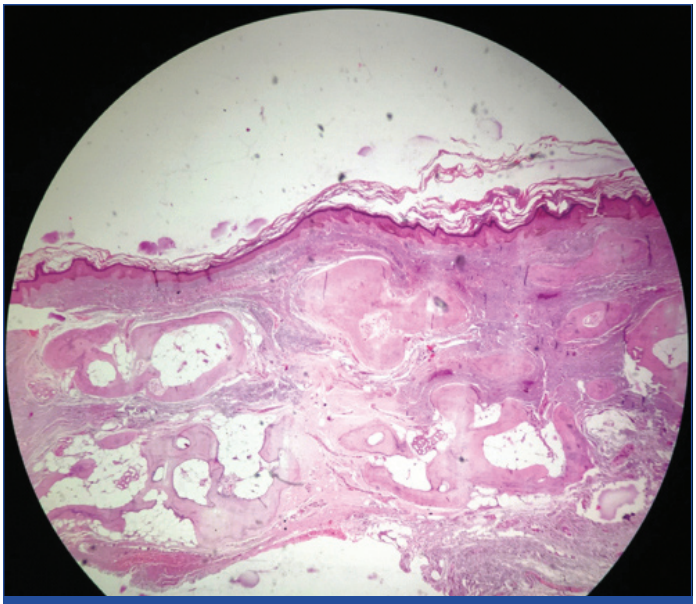
Non-hereditary forms of Heterotopic Ossification (HO) such as myositis ossificans were ruled out on the basis of absence of prior trauma at the sites of lesion or history of surgery and absence of features of arthropathy.

The patient underwent excision of these lesions. Subsequently, her residence shifted post marriage and she was lost to follow-up.

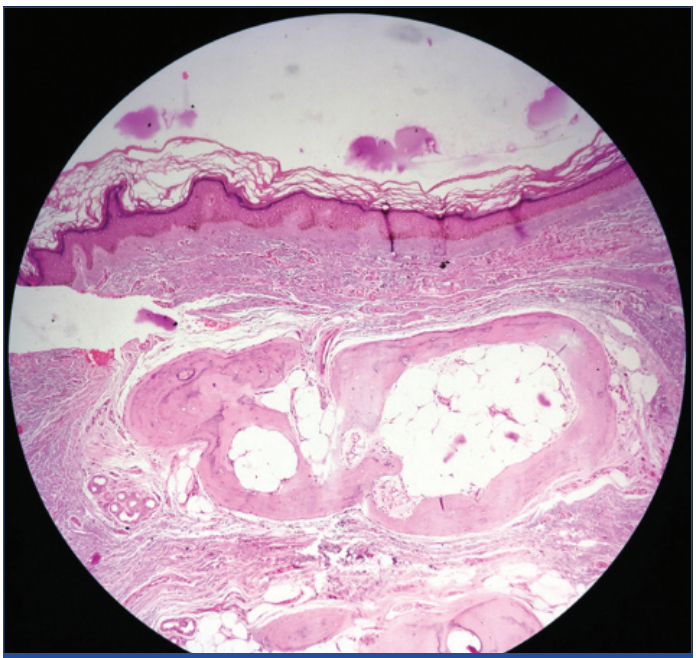
Histopathologic examination of the excised nodules revealed multiple subepidermal spherical foci of ossification [Table/Fig-2,3]. Islands of trabecular bone were seen enclosing spaces containing fat. Various causes of heterotopic ossification observed histologically include myositis ossificans, myositis ossificans progressiva, fibro-osseous-pseudotumour, extraskeletal osteosarcoma, osteomyelitis and pilomatricoma to name a few. These entities were ruled out due to the absence of associated histologic features, fibroblastic proliferation or inflammation [1]. The foci of ossification were well-defined and spherical as opposed to haphazard ossification seen in neoplastic and inflammatory conditions. Thus, the diagnosis of primary osteoma cutis was made.



[Table/Fig-1]: Radiograph right wrist and hand showing multiple discrete radiopaque bony densities indicated by arrows. (Patient consent obtained).



[Table/Fig-2]: Photomicrograph showing multiple subepidermal spherical foci of ossification enclosing fat spaces. (40X, H&E stain) (Patient consent obtained).



[Table/Fig-3]: Photomicrograph showing islands of trabecular bone enclosing fat spaces. (X100, H&E stain) (Patient consent obtained).

DISCUSSION

Osteoma cutis is a rare, benign condition characterised by dermal or subcutaneous bone formation that presents as stony hard nodules [2]. In spite of being benign in nature, these lesions may cause cutaneous deformities [3]. The patient in the present study had approached the Department of Dermatology for mainly cosmetic concerns. Females are more commonly affected especially during the second and third decades [4]. This is in concurrence with the present case where the patient is a 21-year-old girl. However, primary osteoma cutis can present at birth and infancy [5-7].

The most accepted hypothesis regarding the origin of the cell leading to osteoma is fibroblast metaplasia. Another hypothesis states that embryonic mesenchymal cells differentiate into the osteogenic lineage, after being erroneously lodged in the dermis [8,9].

The disease is classified into primary and secondary forms. POC is not associated with any predisposing conditions, whereas SOC, the much more common type, occurs as a result of metabolic conditions that lead to increased blood calcium levels. Scleroderma, pilomatricoma, nevi, basal cell carcinoma, scarsare some of the predisposing conditions [5,10-12]. No predisposing cutaneous condition could be found in the present patient.

The four main syndromes that are associated with the POC are Albright Hereditary Osteodystrophy (AHO), fibrodysplasia of progressive ossification, Progressive Osseous Heteroplasia (POH) and plate-like osteoma cutis [13,14]. These disorders and primary osteoma cutis, share common features of superficial HO in association with inactivating mutations of GNAS [15,16] a negative regulator for osteogenic lineage [17,18].

AHO, commonest of the syndromes associated with POC, is characterised by such physical features as a short stature, round face, obesity, brachydactyly and osteoma cutis. Hypocalcaemia, hyperphosphatemia and an increased serum concentration of Parathyroid Hormone (PTH) are observed in patients with AHO [19]. No facial dysmorphisms or limb malformations could be found in the present patient. Serum estimations of calcium, phosphorus, Vitamin D and parathormone were found to be normal in the present study unlike in AHO. POH is a rare disease in which there is progressive ossification of the dermis in infancy and subcutaneous and deep connective tissue during childhood [20]. Other genetic causes of HO can be excluded on clinical bases. In the present case, absence of radiologic evidence of ossification in the deeper connective tissues helped to rule out POH.

Clinically, the lesions present as single or multiple papules, nodules, plaques or as miliary lesions [21]. The present patient presented with multiple papules on fingers and web spaces. The lesions are bony hard and rarely cause discolouration of the skin which may become yellowish white [22], as in the patient. These nodules sometimes exude fluid from a crateriform opening, perforating osteoma cutis is a very rare condition [2]. Osteoma cutis mostly appears in the periarticular regions such as the scalp, hands and feet, and also face, chest, breast, buttocks, although it can occur anywhere in the body [3,19]. Lesions in the patient were localised in the dorsum of hand and wrist. Gomez Sanchez ME et al., described primary osteoma cutis presenting in the palm [23]. Miliaryosteoma cutis is, when it presents as hardened nodules similar to milia, which particularly affect the face, with a preference for women [8,9]. Less frequently, OC is seen intraorally in the tongue, which is known as osseous choristoma [21].

Management of OC must focus on treating the underlying condition if one exists. As for management of the cutaneous nodules, surgical excision is usually curative without recurrence. Other options include topical tretinoin and dermabrasion [21].

CONCLUSION

Authors herein document an interesting case of osteoma cutis which presented as multiple hard cutaneous nodules in the wrist and hand. Histopathologic examination helped establish a definite diagnosis of osteoma cutis. When faced with a primary osteoma cutis, it is advisable to elicit a thorough clinical history, perform physical examination and investigate for pseudohypoparathyroidism. In this manner, a more accurate diagnosis and appropriate management can be instituted.

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