A Case of Bizarre Parosteal Osteochondromatous Proliferation of the Hand

J L Soon, H C Chang, C S Sim, L C Teoh, C O Low

ABSTRACT

We report a case of Bizarre Parosteal Osteochondromatous Proliferation (BPOP) of the hand that was initially misdiagnosed as an osteochondroma. BPOP, although a benign lesion, behaves aggressively with rapid growth and high risk of local recurrence after local resection. The distinguishing features of BPOP as opposed to an osteochondroma are discussed.

Keywords: Nora's lesion, differential diagnosis, histopathology

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INTRODUCTION

Bizarre parosteal osteochondromatous proliferation (BPOP, Nora's lesion)) is a rare benign lesion that grows rapidly, and recurs in fifty percent of reported cases following resection[1-3]. It has atypical histopathological features that must be distinguished from chondrosarcoma[1,3], low-grade parosteal osteosarcoma[2,3], and conventional osteosarcoma[2,3]. Amongst the benign lesions, florid reactive periostitis, myositis ossificans, periosteal chondroma and osteochondroma should be considered[1,5,6]. We report a case of Nora's lesion that was initially diagnosed as an osteochondroma.

CASE REPORT

A 36-year-old male was seen for a one-year history of a mass on the palmar aspect of the left long finger, located at the level of the metacarpo-phalangeal joint. He worked as a building contractor and had a history of Graves's thyrotoxicosis. There was no history of trauma to his hand. A plain radiograph of the lesion was ordered by the patient's primary physician and the presumptive diagnosis of osteochondroma was made. The mass increased in size but was painless. Subsequently, the patient was referred to our institution for evaluation.

Clinical examination revealed a hard mass, 3 cm in diameter over the palmar aspect of his third metacarpo-phalangeal joint (Fig. 1). It was attached to the proximal phalange of the long finger and the overlying skin was freely mobile. Neuro-vascular examination was unremarkable, and the flexor tendons of the fingers were intact. Radiographic examination revealed a bony mass with well-defined margins taking origin from the base of his left long finger proximal phalanx (Fig. 2a, b).

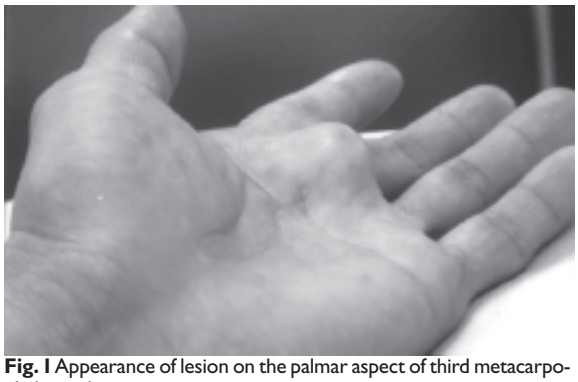

Fig. 1 Appearance of lesion on the palmar aspect of third metacarpo-phalangeal joint.

Computed tomography of the mass was mistakenly reported as an osteochondroma of the left proximal half of the palmar radial aspect of his long finger proximal phalanx. Upon further review, the lack of continuity of the medullary canal of the proximal phalanx and the lesion (Fig. 3) is more characteristic of Nora's lesion[1,2].

Excisional biopsy of the lesion was performed. A dorso-lateral longitudinal incision was made to expose the tumour base, whilst a volar Z-incision over the tumour facilitated its removal. Intra-operatively, a large well-circumscribed pedunculated whitish tumour was found arising from the base of the proximal phalanx. Areas of necrosis were noted on the surface of the lesion. The surrounding structures including the neuro-vascular bundles were easily dissected away from the lesion. The lesion was osteotomised at the base. The gross specimen measured 4 x 3 x 3 cm and had a cartilage cap measuring seven millimetres thick. There was no evidence of malignant change on histological examination, and the diagnosis of osteochondroma was made.

Department of Orthopaedic Surgery
Changi General Hospital
2 Simei Street 3
Singapore 529889

J L Soon, MBBS
Resident

H C Chang, MBBS, FRCSEd (Orth)
Associate Consultant

C O Low, MBBS, FRCS (Glasg), FAMS
Chief and Senior Consultant

Department of Pathology
Changi General Hospital

C S Sim, MBBS, FRCPA
Senior Consultant

Department of Hand Surgery
Singapore General Hospital
Outram Road
Singapore 169608

L C Teoh, MBBS, FRCS (Glasg), FAMS
Head and Senior Consultant

Correspondence to:
H C Chang
Tel: (65) 6850 3571
Fax: (65) 6260 1712
Email: haw_chong_chang@cgh.com.sg

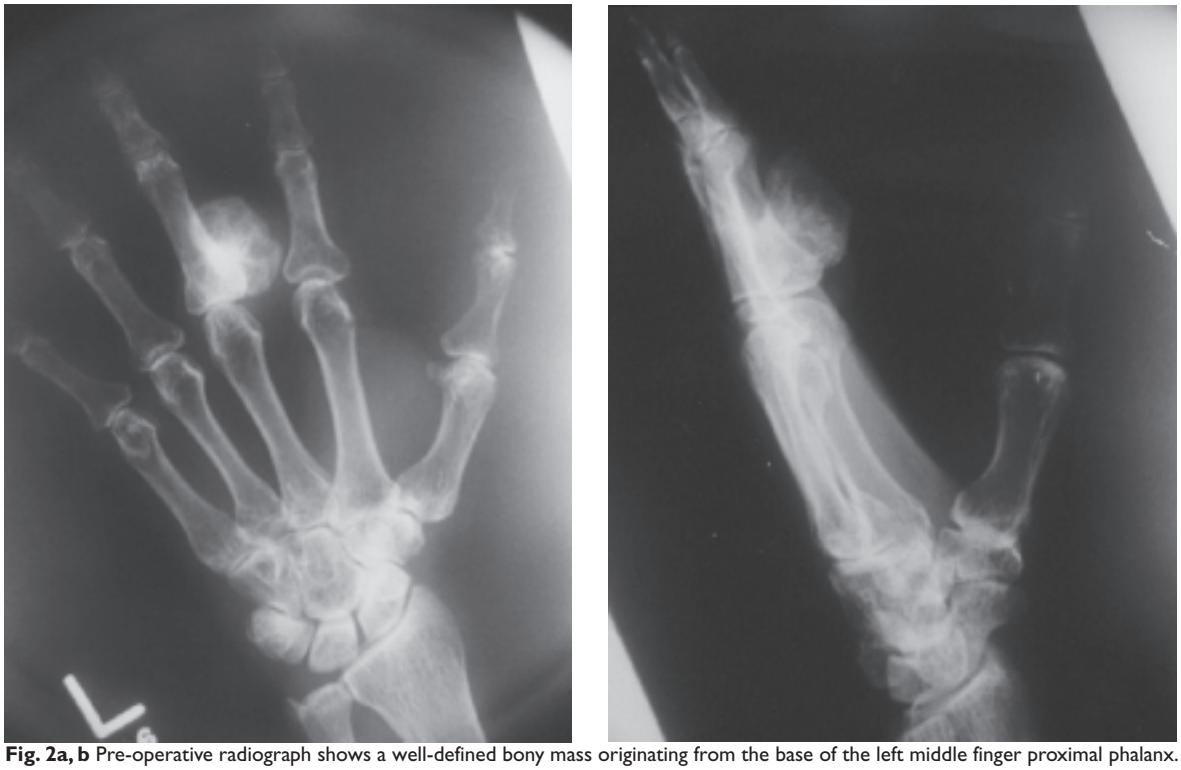

Fig. 2a, b Pre-operative radiograph shows a well-defined bony mass originating from the base of the left middle finger proximal phalanx.

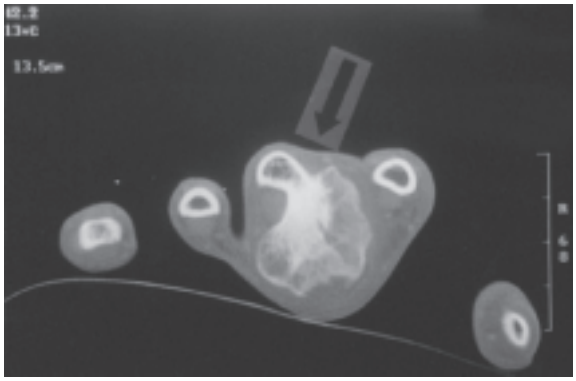

Fig. 3 Axial CT scan shows the lack of continuity of the medullary canal of the proximal phalanx and the lesion, characteristic of Nora's lesion.

At five months post-operative, the patient presented with a recurrent mass in the previous operative site, measuring approximately 3 x 2 cm. Radiographs showed a recurrent tumour from the same area. He underwent a repeat excision.

The radial digital nerve of the long finger was cut during dissection of the scarred tissue, and had to be repaired. The tumour was excised off the base of the proximal phalange and a cuff of normal looking bone was excised as well to minimise the risk of recurrence. The excised specimen was a well circumscribed fibro-myxoid nodule measuring 1.5 x 1.0 x 0.5 cm with focal calcification. Histologically it shows lobulated proliferative cartilage with irregular bone-cartilage interface (Fig. 4a, b) and few binucleated cells. The diagnosis was revised to that of a bizarre parosteal osteochondromatous proliferation (Nora's lesion). It was interesting that when two other pathologists reviewed the histopathology slides of the first operative specimen, they agreed that despite the more irregularly arranged chondrocytes compared to a typical osteochondroma, they would still have labelled it an osteochondroma. The wound and nerve function recovered fully on follow-up, and no tumour recurrence was noted one year later.

DISCUSSION

Bizarre parosteal osteochondromatous proliferation (BPOP), or Nora's lesion was described by Nora et al in 1983[1]. It is a rare lesion commonly affecting the hands and feet of young adults in their 20's and 30's. Males and females are equally affected. It most commonly occurs on the proximal and middle phalanges, metacarpal, and metatarsal bones[2]. BPOP typically presents with a painless or mildly painful mass that grows over a period of weeks to months[1].

In contrast, although osteochondromas can occur in any bone that develops by enchondral ossification, it only affects the small bones of the hand and feet in approximately 10% of cases[5]. Seventy to 80% of solitary osteochondromas occur in children or adolescents younger than 20 years of age, and the growth of the lesion usually ceases with fusion of the adjacent growth plate[5].

The radiographic features of a solitary osteochondroma, arising from the external surface of long bone metaphysis, usually are diagnostic (Table I). The major difficulty is in distinguishing between a benign osteochondroma and a peripheral

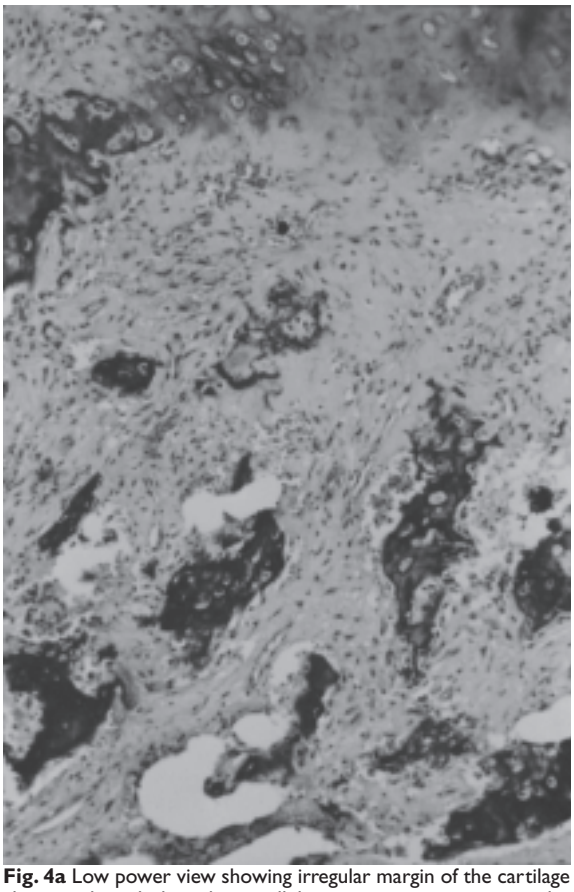

Fig. 4a Low power view showing irregular margin of the cartilage above with underlying hypercellular stroma containing irregular islands of cartilage. (Original magnification HE x 10.)

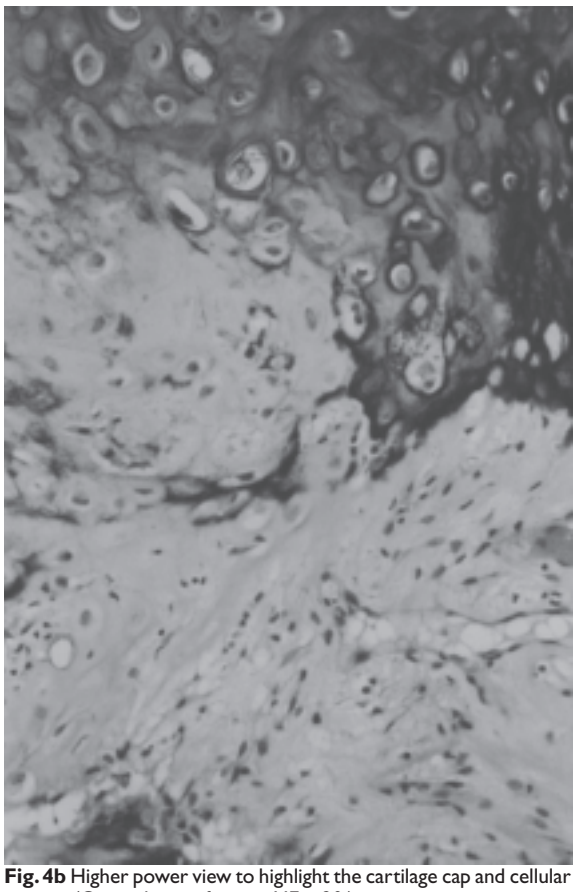

Fig. 4b Higher power view to highlight the cartilage cap and cellular stroma. (Original magnification HE x 20.)

chondrosarcoma in certain cases[5]. Although rare, peripheral chondrosarcoma is the most frequent non-haematological tumour in small bones of the hand[5]. BPOP appears as a well-defined bony mass on plain radiograph[2,3], very much like an osteochondroma except that it may lack the characteristic orientation away from the nearby physis that is seen in the latter[5]. Computed tomography scan helps to distinguish it from osteochondromas by showing the absence of continuity between the cortex and medullary cavity of the bone and the lesion[1,2,5]. The well-formed cartilage cap of osteochondromas, is well demonstrated as high signal intensity on T2-weighted spin echo MR images[5]. A cartilage cap greater than 1-cm thick in adults should also raise the suspicion of chondrosarcomatous transformation of osteochondromas. In contrast to malignant lesions, BPOP exhibits no periosteal reaction, and has normal underlying bone and adjacent soft tissue radiologically.

BPOP has been described as a lesion with a nodular surface covered by glistening cartilage and a distinct blue tint of the bone within on gross examination[2]. It contains disorganised and irregular cartilage with patchy ossification. The periphery of the lesion contains lobulated hypercellular cartilage and fibrous tissue with occasional large chondrocytes. There are uniform osteoblasts on the bony trabeculae. Chondrocytes may be binucleated[1] and proliferation of bizarre (even spindle shaped) fibroblast in the intertrabecular spaces of disorganised bone has been reported[3]. Typically the cartilage lobules show mild atypia and moderate mitotic activity is seen in the spindle cells. This often confuses the diagnosis and the lesion needs to be distinguished from conventional osteosarcoma, low-grade parosteal osteosarcoma, and chondrosarcoma.

Osteochondromas on the other hand, contains a few-millimeters thick chondral cap resembling normal cartilage, beneath which is cancellous bone that is in direct continuity with the underlying bone[5]. The cap however, may be entirely absent in adults, and binucleated chondrocytes may be found in young patients[5]. In contrast to BPOP, osteochondromas have normal "columnation" of the cartilage[1]. Being a rare lesion, the diagnosis of Nora's lesion is challenging for most pathologists as illustrated by this case and others[4].

Although BPOP is known to recur locally in 50% of cases between two months and two years after surgery, it is a benign lesion. Metastatic lesions have never been reported, even in the largest series of 65 patients published by Meneses et al[2]. We report

a case of BPOP that was initially diagnosed as an osteochondroma, and describe the distinguishing features of both lesions. BPOP should be considered when the diagnosis of osteochondroma is made.

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