Case report

Nora's lesion of 2nd metacarpal treated by wide excision, autologous fibular grafting and metacarpophalangeal joint replacement: A rare case report[☆]

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ARTICLE INFO

Article history:

Received 7 July 2017

Received in revised form 1 August 2017

Accepted 3 August 2017

Available online 24 August 2017

Keywords:

Fibular grafting

Metacarpal

Metacarpophalangeal joint replacement

Nora's lesion

ABSTRACT

Bizarre paroxysmal osteochondromatous proliferation (BPOP) is a rare benign neoplastic lesion predominantly affecting small bones of the hand and known for its recurrence after local excision. We describe a rare case of BPOP involving second metacarpal of the dominant hand which was treated using a novel technique. Wide excision and reconstruction with non-vascularised fibular autograft was performed along with metacarpophalangeal joint replacement. At a follow up of two years, there was no evidence of recurrence and patient had good functional outcome. Hence this modality of treatment could be considered in dealing such cases of BPOP involving the small bones of hand.

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1. Introduction

Bizarre paroxysmal osteochondromatous proliferation (BPOP) was first described by Nora and his colleagues in 1983.¹ It is an extremely rare lesion usually involving the bones of hand. There have been fewer than 150 cases of BPOP reported involving various bones.^{2–4} Given the rarity of the condition there is no consensus on its treatment. Majority of the cases have been treated with local excision,⁵ however few authors have suggested wider excision to reduce the chance of recurrence.⁶ We present a new method of treatment in BPOP involving metacarpals of hand. It includes partial excision of metacarpal and reconstruction with fibular grafting and metacarpophalangeal joint replacement.

2. Case report

A fifty-year-old female presented to our outpatient department with swelling in the right hand (dominant) of one year duration. There was gradual increase in the size of swelling from size of pea to approximately that of a lemon. There was no history of trauma or fever or any other complaint. It was not associated with pain. On examination a diffuse bony hard swelling of size 4 × 4 cm was localised to the dorsal aspect of the index metacarpal with no tenderness (Fig. 1). There was no distal neurovascular deficit. Rest of the clinical examination was unremarkable.

Radiograph of the right hand revealed a well-defined heterogeneous radiolucent lesion appearing to be arising from the surface of 2nd metacarpal with matrix calcification (Fig. 2). Magnetic resonance imaging (MRI) and computed tomography (CT) of the hand showed a well-defined lytic destructive lesion arising from the extensor aspect of the second metacarpal contiguous with the medullary cavity of the underlying bone and internal lobulations suggestive of chondroid matrix (Figs. 3 and 4). Core biopsy was done from the lesion which revealed tumoral proliferation with osseous, cartilaginous and fibrous components suggestive of BPOP (Fig. 5). The patient was then planned for wide excision of the lesion and reconstruction with fibular grafting and metacarpophalangeal joint arthroplasty using NeuFlex prosthesis (DePuy Orthopaedics, Warsaw, IN, USA).

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Fig. 1. Clinical photograph of the lesion. A diffuse bony hard swelling of size 4×4 cm was localised to the dorsal aspect of the index metacarpal of the right hand.

Under general anaesthesia and tourniquet control, a straight midline skin incision was given over the dorsum of second (index) metacarpal extending upto proximal phalanx. The extensor tendon was retracted to one side. The mass was dissected from all sides taking care not to breach the pseudocapsule. Metacarpal osteotomy was done two centimetres proximal to the lesion and the mass was removed along with the second metacarpophalangeal joint. After excision of the mass, the wound was thoroughly washed with normal saline. Appropriate sized (five cm) non-vascularised fibular

graft was harvested from ipsilateral leg. The fibular graft was then shaped and trimmed to be placed over the defect in the second metacarpal. The articular cartilage was then removed from the proximal phalanx of second digit with the help of micro-oscillating saw. The medullary canal was opened with the help of micro rasp. Sequential broaching was carried out in both proximal phalanx and harvested fibular graft. The trial silicone prosthesis of size one was then inserted in the proximal phalanx and the fibular graft. The fibular graft was fixed to remaining base of metacarpal with the help of 2.4 mm locking compression condylar plate (AO, Synthes). After checking stability of the construct, the trial prosthesis was removed and definitive prosthesis of size 1 was then inserted and the joint reduced. After thorough washing of wound, it was closed in layers. The finger was splinted for two weeks and subsequently active range of motion exercises were advised to the patient. There was no post-operative complication (Fig. 6).

The excised mass was sent for histo-pathological examination. On microscopy, there was fibrocartilaginous tissue with high cellularity; small chondrocytes were scattered in myxoid stroma. There were areas of immature bony trabeculae with high osteoblastic activity. The spindle cells were arranged loosely along the trabeculae. No cellular atypia or mitoses was seen. The above-mentioned findings confirmed the final diagnosis of BPOP of the second metacarpal. At a follow up of two years, there was no evidence of recurrence. The patient was able to write and pinch with good functional outcome (Fig. 7).

3. Discussion

BPOP in hand presents as an osseous exophytic growth. It affects men and women of all age groups with equal frequency.⁶ Patients usually present with a non-tender and firm mass over the dorsum of hand. Sometimes joint motion may be limited due to its location at the end of the joint. Its aetiology and pathophysiology are not known. Some authors have proposed traumatic event in its aetiology^{3,7} while others have refuted the same.⁸ Yuen et al. in their unitary hypothesis have proposed that BPOP along with florid

Fig. 2. Plain radiograph of the right hand. A well defined heterogenous radiolucent lesion appear to be arising from the surface of 2nd metacarpal with matrix calcification.

Fig. 3. NCCT scan with 3D reconstruction of the right hand. A well defined lytic destructive lesion arising from the extensor aspect of the 2nd metacarpal contiguous with the medullary cavity of the underlying bone.

Fig. 4. MRI of the right hand. T2 Sagittal and axial MRI images showing well defined hyperintense mass lesion contiguous with the medullary cavity.

reactive periosteitis and turret exostosis form a continuum and each represents a different stage in proliferative reaction of periosteum.⁹ A traumatic event leads to subperiosteal haemorrhagic proliferation and formation of florid reactive periosteitis which is followed by BPOP and turret exostosis in the final stage. However in our case there was no history of trauma.

Histologically BPOP has cartilage, bone and fibrous tissue in varying amounts. There may be a cartilaginous cap and sometimes the cartilage may arrange in the form of lobules which are separated by fibrous tissue. The cartilage may contain binucleated

chondrocytes but there is no hyperchromasia and cytological atypia. The bone formed in cartilaginous lobules stains dark blue especially at the cartilage interfaces. The spindle cells present in intr trabecular spaces do not show atypical cells.^{3,6}

On the basis of the proliferative osteochondromatous composition and location of the lesion, the differential diagnosis included osteochondroma, parosteal osteosarcoma, myositis ossificans and chondrosarcoma. All cases of BPOP must be distinguished from these conditions in order to plan further management. Osteochondromas are among the most common benign bone tumours;

Fig. 5. Histopathological microphotographs of the lesion. a) Microscopic view showing bone trabecula associated with fibrous tissue (40 $\times$ magnification), b) Microscopic view showing a tumoral proliferation with osseous, cartilaginous and fibrous components (40 $\times$ magnification).

Fig. 6. Post-operative radiograph. Anteroposterior and lateral views of the right hand showing fibular graft fixed with 2.4 mm locking compression condylar plate and the metacarpophalangeal joint replacement.

Fig. 7. Post-operative clinical photographs of the patient. The patient can actively make a fist and do pinching action with the index finger.

however they have preponderance for the long bones. On histological examination, osteochondromas do not show atypical cells and there is regular chondrocyte alignment. Parosteal osteosarcoma can be differentiated from BPOP by presence of mature bony trabeculae, atypical fibrocytes, spindle cell proliferation and presence of cartilage over the surface or deep inside the mass. Myositis ossificans has distinct history of trauma and centripetal pattern of ossification. Chondrosarcoma shows myxoid matrix with hypercellularity and marked anaplasia.

BPOP is a locally aggressive lesion and intralesional excision may lead to its recurrence. A recurrence rate of upto 50% has been reported with this lesion.³ The formal excision of pseudocapsule covering the cartilage cap and abnormal periosteum and adjacent cortical area decreases the recurrence rate.¹⁰ The treatment modality may vary from primary amputation to excision and reconstruction with fibular struts depending on the site of involvement.¹⁰ In our case the metacarpal lesion was close to the metacarpophalangeal joint and hence decision to excise the distal end of metacarpal shaft and MCP joint was taken. This would have led to a large void over the distal end of second metacarpal and an absent MCP joint. So we decided to reconstruct the void with non-vascularised fibula and MCP joint replacement. The patient had good functional results at the end of two years. In many cases where wide excision is carried out, there is instability due to inadequate reconstruction. On the other hand, in those cases where reconstruction is adequate, there may be stiffness at the small joints due to long period of immobilisation required as kirschner's wire are commonly used for holding the fibular graft.¹⁰ Range of movements can be initiated at an early stage in case of stable reconstruction with miniplates and metacarpophalangeal joint replacement and hence good results can be obtained. This modality of treatment has never been described to the best of our knowledge in literature in the treatment of BPOP.

Funding source

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

Conflicts of interest

None.

Acknowledgements

None.

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