Case Reports

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Nora's lesion of the distal ulna: a case report

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Abstract

Nora's lesion, also known as bizarre parosteal osteochondromatous proliferation (BPOP), is a very rare benign lesion with few published cases. BPOP is more common in adults during the second to third decades of life, and usually occurs on the hands and feet. Radiologically, it appears as a calcified mass attached to the bone cortex that grows rapidly and that recurs easily following resection. Aggressive features on imaging and confusing histopathological findings usually result in misdiagnosis or mistreatment. Herein, we present a case of a rare bony tumour involving the distal ulna presenting as a painless growing mass. An excisional biopsy with clear margins was performed without disturbing the ulnar nerve and arteries. There was no recurrent mass or calcified lesion 1 year after surgery. Based on its rarity and difficult diagnosis, BPOP should be considered in the differential diagnosis of a painless mass in the distal ulnar region. Careful follow-up after surgery is essential, even without lesion recurrence.

Keywords

Nora's lesion, bizarre parosteal osteochondromatous proliferation, distal ulna, case report, calcified mass, painless mass, excision

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Background

Nora's lesion, also known as bizarre parosteal osteochondromatous proliferation (BPOP), was first described by Nora et. al in 1983.[1] BPOP is a very rare benign lesion rarely reported in the literature. This lesion is more common in adults during the second to third decade of life and usually occurs on the hands and feet.[2–6] BPOP lesions grow rapidly and often recur after

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resection. Radiologically, the lesion appears as a calcified mass attached to the bone cortex and can mimic an exostosis, osteochondroma or malignancy. Confusing histopathological results and aggressive features on imaging usually result in misdiagnosis or mistreatment. In this report, we present a case of successful treatment of a rare bony tumour involving the distal ulna.

Case presentation

The patient was a 48-year-old man with hypertension that was inconsistently controlled with medication. He visited our clinic because of a growing mass on the volar side of his left wrist that was present for 1 year. He denied a history of trauma or a remarkable family history. According to the patient, the mass had increased in size but was not associated with pain or numbness. There was no skin discolouration, and the neurovascular examination was normal. Full range of motion of the left wrist (flexion, extension, supination and pronation) and fingers (flexion and extension) was intact. Radiographs of the left wrist showed a calcified, ill-defined tumour on the volar side of the distal ulna (Figures 1 and 2), and magnetic resonance imaging (MRI) revealed an ossified mass ($3.0 \times 1.8 \times 1.3$ cm) on the volar ulnar side, with multiple calcified areas. However, the ulnar artery and nerve were not involved with the tumour, and there was no bony connection to the distal ulna (Figures 3 and 4).

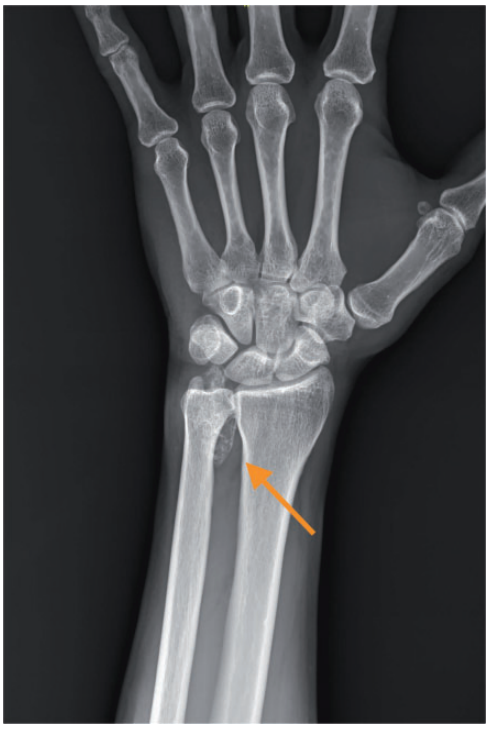

Figure 1. Pre-operative left wrist anteroposterior (AP) and lateral (LAT) views showing a calcified, ill-defined tumour on the volar side of the left ulna.

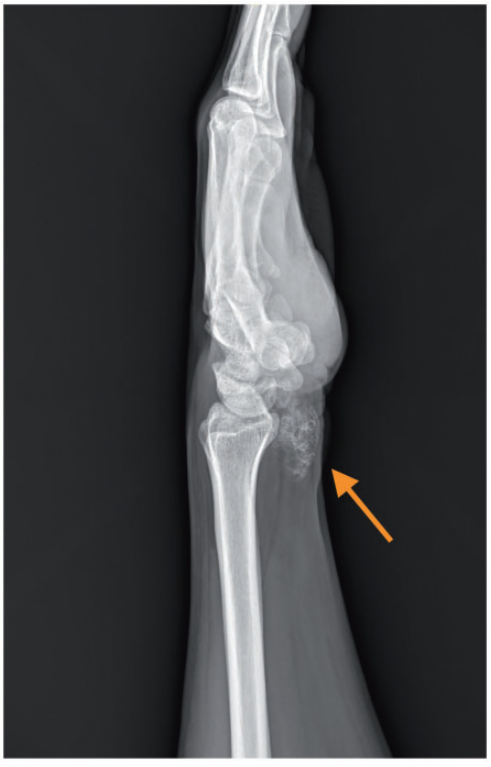

Figure 2. Pre-operative left wrist anteroposterior (AP) and lateral (LAT) views showing a calcified, ill-defined tumour on the volar side of the left ulna.

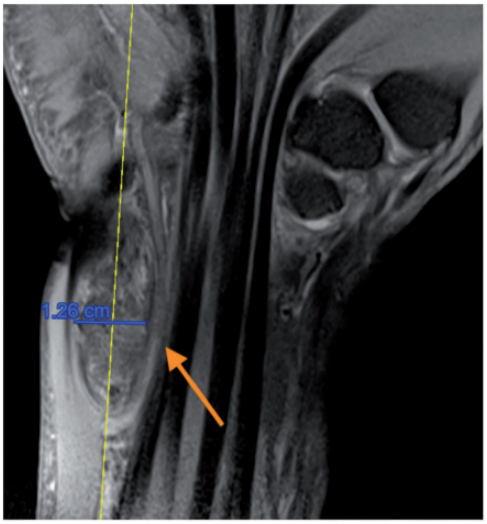

Figure 3. Pre-operative magnetic resonance (MRI), proton density-weighted image with fat suppression phase showing a left distal ulnar (wrist) volar ossified mass. Multiple calcified spots are visible within the tumour, which measured $3.0 \times 1.8 \times 1.3$ cm. The volar cortex of the ulna, and the ulnar artery and nerve were intact.

Because of cosmetic concerns and flexion limitation associated with the enlarged mass, the patient consented to surgical resection for excisional biopsy. A volar approach was adopted, and a pseudo-capsule surrounding the mass was found (Figure 5). We excised the mass completely and cautiously, leaving no gross remnant at the lesion site (Figure 6). We used 95% alcohol to flush the surgical field, and the wound was closed. The postoperative condition was good, and there were no motor or sensory impairments. Pathological reports showed mature osseous tissue mixed with adipose tissue and the presence of a cartilage cap. The chondroid tissue showed mild nuclear atypia, and BPOP (Nora's lesion) was observed (Figure 7 and 8). Immunohistochemistry for mouse double minute 2 (MDM2) and cyclin-dependent kinase (CDK)4 proteins was not performed because parosteal

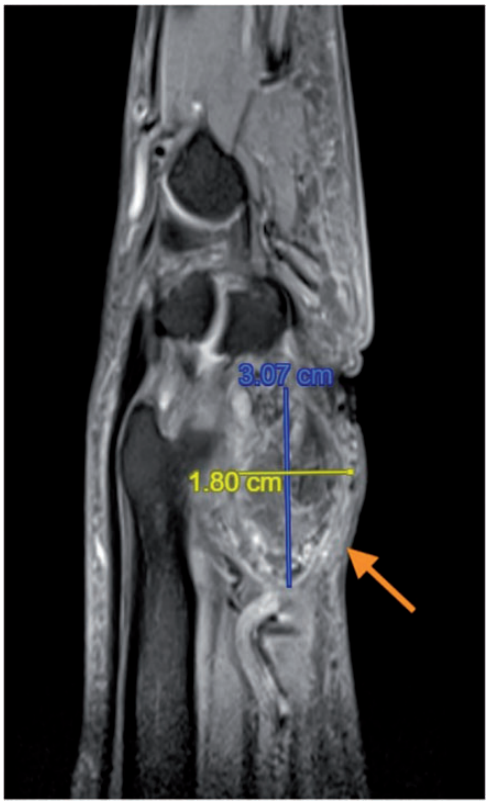

Figure 4. Pre-operative magnetic resonance (MRI), proton density-weighted image with fat suppression phase showing a left distal ulnar (wrist) volar ossified mass. Multiple calcified spots are visible within the tumour, which measured $3.0 \times 1.8 \times 1.3$ cm. The volar cortex of the ulna, and the ulnar artery and nerve were intact.

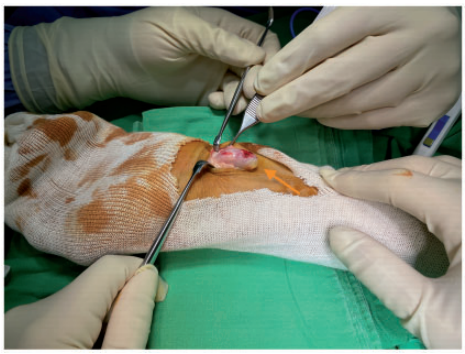

Figure 5. The well-encapsulated volar ulnar mass was resected.

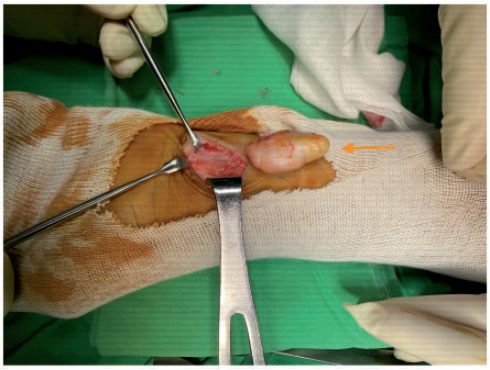

Figure 6. The tumour was completely removed with an intact capsule.

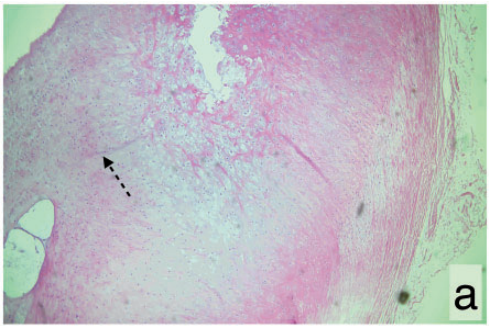

Figure 7. Histological findings in our case. Low magnification (×40) haematoxylin and eosin staining (a and b) showing mature osseous tissue mixed with adipose tissue and a highly cellular cartilaginous cap (long arrows in b) with enlarged chondrocytes, mild atypia and irregular ossification. There are also interspersed regions of fibrovascular stroma with spindle cells (dashed arrow in a).

osteosarcoma was included in the differential diagnosis, and this tumour is usually negative for these markers. Radiography and MRI 1 year after surgery revealed no recurrent tumour or calcification in the left wrist (Figures 9 and 10). The range of motion was complete, and the patient was satisfied with the results of the operation. The reporting of this study conforms to the CARE guidelines.[7]

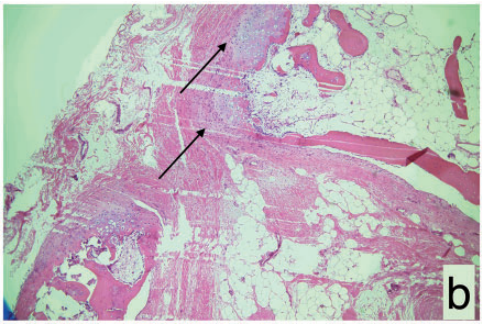

Figure 8. Histological findings in our case. Low magnification (×40) haematoxylin and eosin staining (a and b) showing mature osseous tissue mixed with adipose tissue and a highly cellular cartilaginous cap (long arrows in b) with enlarged chondrocytes, mild atypia and irregular ossification. There are also interspersed regions of fibrovascular stroma with spindle cells (dashed arrow in a).

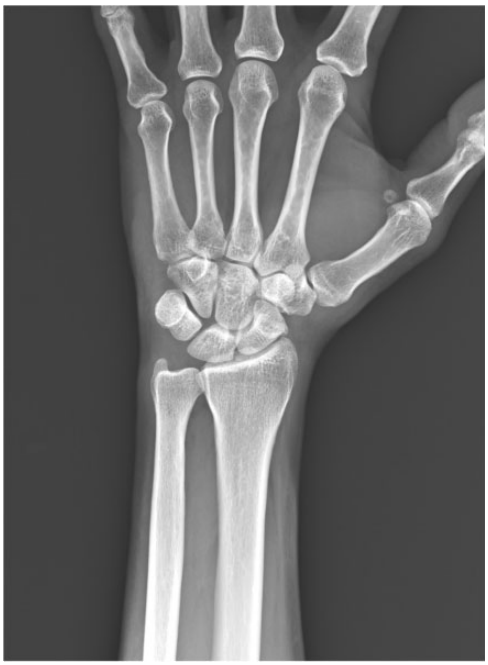

Figure 9. Post-operative X-ray images of the left wrist, anteroposterior (AP) and lateral (LAT) views, showing no remnant mass or calcified areas.

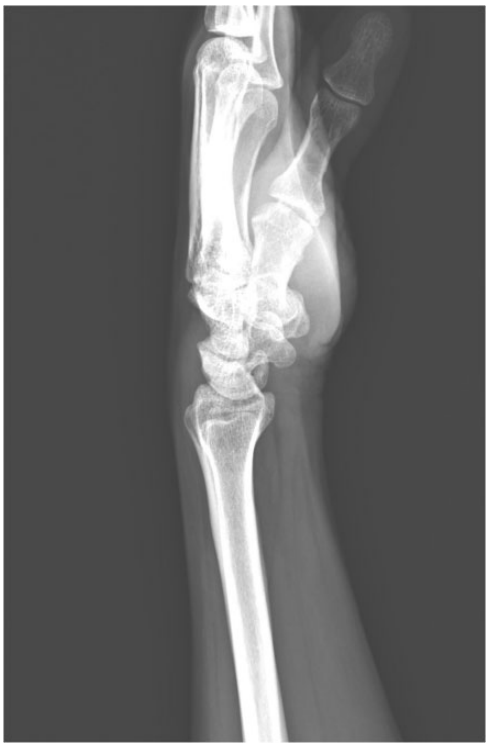

Figure 10. Post-operative X-ray images of the left wrist, anteroposterior (AP) and lateral (LAT) views, showing no remnant mass or calcified areas.

Discussion and conclusions

BPOP is a rare, benign, reactive mineralising mesenchymal lesion, with few reported cases. There are no sex differences, and BPOP is often found in individuals in their 20s and 30s.[1,2,4,5] The most common presentation is a growing mass arising from the metacarpal or metatarsal bone, with few cases described in the long bones.[1,2,5,8] Occasionally, the lesion may be slightly painful, mostly due to the mass effect and decreased range of motion; thus, malignancy is also suspected.[8] Our patient presented with painless swelling and no history of trauma. However, in some instances of BPOP, there can be a history of trauma.

The diagnosis of Nora's lesion is reached using a combination of radiology and, most importantly, histological findings. Radiologically, the lesion appears as a well-demarcated, pedunculated tumour arising from the cortical surface of the underlying bone, with an intact surface and without a periosteal reaction.[2,4] On computed tomography, Nora's lesions show no continuity with the medullary cavity, while osteochondromas maintain continuity. On MRI, Nora's lesions usually display low signal intensity on T1-weighted sequences and high signal intensity on T2-weighted images, with a higher signal intensity on the periphery. The lesion is not enhanced by gadolinium contrast, suggesting cartilage cell proliferation.[5] Careful differentiation during the pathological examination must be made between malignant and benign lesions, especially under a suspicion of malignancy. Histopathological features typical of BPOP comprise a hypercellular cartilaginous cap with regions of incomplete endochondral ossification, densely mineralised cartilaginous tissue/bony trabecula (basophilic bone or blue bone) and interspersed fibrovascular stroma without cytologic atypia.[9]

Diagnosis can be difficult owing to the varied appearances of the lesion at the different disease stages. In addition, BPOP can be easily confused with benign lesions, such as florid reactive periostitis, myositis ossificans, periosteal chondroma and osteochondroma,[3] or malignant lesions, such as chondrosarcoma,[2,10] parosteal osteosarcoma[1,10] and conventional osteosarcoma.[1,10] Benign venous malformations can be ruled out if they are superficial. The high recurrence rate of BPOP may lead to a misdiagnosis of malignancy, and the lesion might be mistaken for osteochondroma because of its surface location and cartilaginous component. Osteochondroma also occurs on the surface of bones but has a distinctive histology showing characteristic endochondral ossification. Osteochondromas show cortical and medullary continuity with the

underlying bone, in contrast to BPOP, where there is usually no medullary involvement on imaging. The absence of other associated changes in the underlying bone helps distinguish BPOP from malignant bone tumours, such as osteosarcoma. For soft tissue tumours, myositis ossificans and periosteal chondroma may also be considered in the radiographic differential diagnosis owing to the similar calcified appearance to BPOP.[11] However, myositis ossificans is a soft tissue mass in which bone formation takes place in muscle or soft tissue; calcification may develop in the intermediate stage. These lesions are separated from the bone by a radiolucent cleft and are often associated with a periosteal reaction in the underlying bone. For periosteal chondroma, the typical radiographic appearance is a calcified chondroid matrix with scalloping and slightly overhanging cortical edges of adjacent bone. In addition, periosteal chondroma tends to lack associated medullary bone or soft tissue oedema.[11] Histologically, myositis ossificans, periosteal chondroma and florid reactive periostitis lack cellular pleomorphism and atypical mitoses. The combination of location and histological findings may lead to a BPOP diagnosis.

The natural evolution of BPOP involves a parosteal soft tissue swelling or mass, sometimes with tiny calcifications, as the first presentation. The second stage is observed as parosteal, flame-like calcifications, while the last stage is a mature osteophytic bony lesion, which is well-demarcated and sessile and arises from the cortical surface of the underlying bone without disruption of the native architecture.[5]

The cause of Nora's lesions remains unclear. One explanation involves the reparative process following trauma to the periosteum, as noted in 30% of the cases in the series by Meneses et al.[2,12] BPOP has also been considered part of the spectrum between florid periostitis and turret exostosis,[5] with each representing a different stage of the proliferative periosteal process. Other hypotheses involve a neoplastic process theory.[13] This hypothesis is supported by a high and often early recurrence rate. Chromosomal anomalies associated with BPOP are t(1; 17) (q32; q21) and t(1; 17) (q42; q23).[4]

Generally, observation alone is considered adequate for asymptomatic Nora's lesions, but excision is indicated for patients with pain or functional impairment.[14] Resection of the lesion capsule and decortication of the underlying cortical bone are important in reducing recurrence rates.[10,13] If the large size or surgical field may result in bone instability, reconstructive procedures are necessary. Wide resection has not been advocated because of the benign nature of Nora's lesion. Despite appropriate treatment, Nora's lesion has a high recurrence rate of 29% to 55% within the first 2 years following initial treatment.[5]

To the best of our best knowledge, there are fewer than five reported cases of Nora's lesion affecting the ulna.[14–16] Rybak et al. reported a 16-year-old girl with a Nora's lesion of the distal ulna. Index surgery was performed uneventfully, but recurrence was noted 3 years later.[16] Matsui et al. reported a 58-year-old Japanese woman with Nora's lesion at the left distal ulna.[14] Complete excision was performed through a volar approach without recurrence after 2 years of follow-up. Washington et al. reported a 22-year-old man with a right wrist mass after trauma.[15] BPOP was suspected with MRI and was confirmed pathologically. In our case, definitive excision with gross negative margins was performed, then the surgical field was irrigated with 95% alcohol, which has been proven to reduce the local recurrence of giant cell tumours.[17] Our patient developed no gross or radiological local recurrence, and the range of motion in his wrist was complete and without

discomfort at the 1-year follow-up. Serial follow-up is scheduled for our patient because recurrence is reported to occur from 10 to 120 months (average: 49 months) after surgery.[18]

Nora's lesion is a rare disease that is extremely uncommon in the distal ulna, and it has a high local recurrence rate after surgical resection. We reported a case of Nora's lesion in the distal ulna that was suspected based on preoperative imaging studies, which led to *en bloc* lesion resection and 95% alcohol irrigation to reduce the possibility of recurrence. The final diagnosis was confirmed according to detailed histopathological findings. One year after surgery, the patient had no subsequent pain or recurrent mass, and he had perfect range of motion. However, given the high local recurrence of this type of lesion, careful follow-up is essential.

Availability of data and materials

Not applicable.

Author contributions

CH Lin attended the surgery, provided the patient's primary care, wrote the manuscript and discussed the findings.
K Wu performed the surgery, oversaw the patient's care, revised the manuscript and provided useful suggestions.
All authors have read and approved the manuscript.

Declaration of conflicting interest

The authors declare that there is no conflict of interest.

Ethics statement

This study was approved by the Far Eastern Memorial Hospital Ethical Review Board. The person described in this report provided written informed consent for participation and for the publication of the related images and data.

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