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We describe 22 cases of bizarre parosteal osteochondromatous proliferation, or Nora’s lesion. These are surface-based osteocartilaginous lesions typically affecting the hands and feet. All patients were identified from the records of a regional bone tumour unit and were treated between 1985 and 2009. Nine lesions involved the metacarpals, seven the metatarsals, and five from long bones (radius, ulna, tibia, and femur in two). The mean age of the patients was 31.8 years (6 to 66), with 14 men and eight women. Diagnosis was based on the radiological and histological features. The initial surgical treatment was excision in 21 cases and amputation of a toe in one. The mean follow-up was for 32 months (12 to 162). Recurrence occurred in six patients (27.3%), with a mean time to recurrence of 49 months (10 to 120). Two of the eight patients with complete resection margins developed a recurrence (25.0%), compared with four of 14 with a marginal or incomplete resection (28.6%).

Given the potential surgical morbidity inherent in resection, our data suggest that there may be a role for a relatively tissue-conserving approach to the excision of these lesions.

Bizarre parosteal osteochondromatous proliferation (BPOP), or Nora’s lesion, is a reactive mesenchymal lesion of bone characterised by an exophytic growth comprising bone, fibrous tissue and cartilage.1 Nora, Dahlin and Beabout2 initially described 35 cases affecting the metacarpals, metatarsals or phalanges; others have since reported the condition at numerous other sites.3-9 Debate about the underlying pathophysiology continues. Although trauma has been proposed as an aetiological factor,3,10,11 this has been refuted elsewhere.12,13 Some authors have recently attempted to reclassify BPOP as a neoplastic process14,15 and a specific genetic translocation has been identified.16

There are conflicting views about the radiological diagnosis. BPOP is classically a broad-based calcified lesion attached to the cortex.17 Lack of medullary continuity differentiates it from osteochondroma.18 However, Rybak et al19 refute this, describing instances of corticomedullary continuity in histologically proven BPOP. The definitive diagnosis therefore remains histological,20 with features suggesting a reactive or reparative lesion.21 BPOP is considered to be benign, despite a growth pattern which occasionally mimics a more aggressive neoplastic process;14 there is no suggestion that it is a premalignant condition.22 However, difficulty in making a definitive clinical or radiological diagnosis means that excision is considered mandatory, even in patients without symptoms from local pressure.23 Some authors advocate local excision;23 others advise wider excision with a view to reducing the likelihood of recurrence.20,24 Because of this ongoing difference of opinion, we decided to review our experience of patients with this rare condition.

Patients and Methods
A retrospective search was undertaken of our regional bone tumour unit, which dates back to January 1985. We initially evaluated the notes of all patients identified. Cases referred for an opinion on treatment, or histological assessment only, were excluded from the study. Only 22 histologically confirmed cases of BPOP, treated at our hospital, were included in the final data. There were 14 men and eight women, with a mean age of 31.8 years (6 to 66).

Nine lesions involved metacarpals (40.9%) and seven the metatarsals (31.8%); one affected a sesamoid in the foot (4.5%), and five occurred in long bones (22.7%): the proximal radius, distal ulna, and lateral tibial condyle in one case each, and the femur in two (distal femoral metaphysis and medial femoral condyle, respectively). The mode of diagnosis, definitive treatment, subsequent follow-up and recurrence rates of all patients were reviewed.
Data were analysed using Fisher’s exact test. A p-value < 0.05 was considered to represent statistical significance.

Results
Clinical presentation. Five lesions presented with a painless swelling, four with a painful swelling and three with pain alone. One, affecting the proximal radius, presented with a painless limited range of movement, and three presented with a painless swelling following trauma. The remaining six had no documented presenting features. The diagnosis was based on the combined radiological and histological features, as outlined below. In 20 cases a formal excision biopsy was undertaken immediately after radiological investigation, and in two a CT-guided needle biopsy was undertaken before the lesion was excised. In seven patients it was felt that a firm diagnosis could be made on clinical and radiological grounds alone, although all subsequently underwent surgery.

Radiological features. On plain radiographs each lesion presented as an irregular, variably calcified mass arising from the surface of the bone but not breaching the cortex (Fig. 1). A CT scan was performed in four cases. In these cases, CT confirmed a formal cortical and medullary pattern. In 21 cases no medullary or cortical continuity between the lesion and underlying bone was seen. In one there was a small area of medullary continuity between the major osseous component of the lesion and the underlying phalanx. MRI, performed in five patients, showed mild oedema of soft tissue around the lesion.

Management. Surgical treatment was by excision in 17 patients, supplemented in two cases by curettage, ‘shave’ excision in three, ‘shark-bite’ excision in one, and in one primary amputation of a toe, owing to the size of the lesion. The shark-bite excision (taking a margin of normal bone around the lesion) was performed in the distal ulna, because of pre-operative concern about a possible diagnosis of osteoblastoma. This patient made a good recovery and remains free of recurrence after 21 months. The remaining lesions in long bones were treated by shave excision (tibial condyle) and marginal excision (proximal radius, distal femur and femoral condyle). In one case, in which a large lesion involved the volar aspect of the fourth metacarpal, metacarpal excision was combined with reconstruction using a fibular strut graft (Fig. 2). After 16 months the patient remained disease-free, with no loss of function. The mean time from the onset of symptoms to surgery was 21 months (1 to 120), although this figure is limited by the accuracy of patient recall. The mean follow-up after surgery was 32 months (16 to 162).

Histological findings. Macroscopically, all lesions were exophytic with a smooth cartilaginous cap. Microscopic
examination revealed anastomosing trabeculae of woven bone, often maturing into lamellar bone covered by a cap of variably calcified cartilage containing chondrocytes, and merging with periosteum-like fibrous tissue at the margins (Fig. 3). The trabeculae had a characteristic deep blue basophilia on staining with haematoxylin and eosin. One case showed the unusual feature of medullary involvement; another showed uncharacteristically poor demarcation from surrounding soft tissues. No lesion showed evidence of malignancy or cellular atypia.

Rates of recurrence. Recurrence occurred in six patients (27.3%). The mean time from initial treatment to the diagnosis of recurrence was 49 months (10 to 120). One patient had five clinically distinct recurrences over a period of 13 years, culminating in ray amputation (counted as one recurrence case for the purposes of data analysis). Of the 14 patients who had a marginal or incomplete resection, recurrence was seen in four (28.6%), compared with two of eight (25.0%) whose resection had been complete. This difference was not statistically significant (p = 0.376).

Discussion

BPOP is a rare condition. To our knowledge, the cases presented here represent 10% of all those reported to date. Its recurrence rate of 28% is comparable to that of Dhondt et al.,25 with others describing rates between 20%26 and 55%.3 This is a small, retrospective case series, precluding formal statistical comparison between the complete and incomplete excision groups. Nevertheless, this review has highlighted aspects of the management of this rare condition, and our experience seems to correlate broadly with that of others. Most agree that the diagnosis must be histological; this is supported by our series, with significant doubt over the diagnosis in 14 cases until confirmed by the histology. Although we have included one histologically confirmed case with corticomedullary continuity, which is uncharacteristic of BPOP, this has been previously described.19

Some groups propose that formal excision of both the overlying pseudocapsule and the periosteum deep to the lesion may limit recurrence.12,18,20 However, others recommend that wide excision, which potentially carries the risk of surgical morbidity, should not automatically be considered as first-line treatment.1,21 Interestingly, our data do not necessarily suggest that wider initial excision reduces recurrence. There may be an argument for undertaking a relatively limited initial resection to minimise morbidity, although this decision must be based on the absence of any suspicion of malignancy. We are currently formulating local management guidelines for BPOP, and agree with previous proposals that patients with this condition should be treated and undergo long-term follow-up in specialist tumour centres.1

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References