Painless wrist lump

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Diagnosis

Parosteal osteosarcoma of the distal radius

Discussion

Radiographs showed an extra-osseous, well-defined ossified mass arising from the volar, distal radial metadiaphysis with a lobular ‘cauliflower-like’ morphology. There was adjacent cortical thickening but no overt cortical destruction or aggressive periosteal reaction (Fig. 1). CT showed stippled mineralisation extending from the volar aspect of the cortex with medullary continuity (Fig. 2). The lesion had a broad base and was more ossified centrally than peripherally, with no appreciable soft tissue mass or cortical destruction. MRI demonstrated a dome-shaped mass measuring 14 × 12 × 10 mm arising from the volar radial cortex (Fig. 3). This was of heterogeneous low T1W SI and heterogenous intermediate-to-low T2W SI centrally with thin peripheral hyperintensity suggestive of a poorly defined cartilage cap. Involvement of the cortex was optimally demonstrated on the post-contrast fat-suppressed T1W TSE image sequences with minor underlying marrow abnormality suggestive of early medullary involvement. The lesion was considered likely benign, with bizarre parosteal osteochondromatous proliferation (BPOP)

the initial diagnosis. Excision biopsy was therefore performed. Histopathology sections showed a tumour composed of anastomosing trabeculae of lamellar and woven bone with intervening mildly atypical fibroblastic spindle cells arranged in vague fascicles and embedded in a densely collagenous stroma. Occasional mitoses were identified (3/10HPF) with no atypical forms present. The tumour also had an incomplete cartilaginous cap with no high-grade cytological atypia identified (Fig. 4). Fluorescence in situ hybridisation showed *MDM2* amplification and therefore the tumour showed classical features of a low-grade parosteal osteosarcoma.

Repeat MRI obtained 4 weeks post-operatively (Fig. 5) demonstrated residual surface and medullary tumour. Completion staging with whole-body MRI and CT chest showed no metastases, and the patient subsequently underwent a ‘shark-bite’ re-excision 6 weeks after the original operation; the resection specimen is demonstrated in Fig. 6.

Parosteal osteosarcoma is the commonest surface osteosarcoma, accounting for 4% of osteosarcomas [1]. Unlike conventional osteosarcoma, there is a slight predilection for females and an older age range, with a median age at presentation in the 4th decade [1, 2]. It is a malignant but low-grade tumour with dedifferentiation being observed in approximately 25% of cases, most commonly to high-grade surface osteosarcoma or spindle cell sarcoma [3]. Medullary involvement on cross-sectional imaging is reported to correlate with the degree of dedifferentiation [2, 4], although reactive remodelling of underlying bone may cause confusing imaging appearances [5]. Survival rates are higher than for conventional osteosarcoma [1].

The femur is the commonest site affected (approximately two-thirds of cases and usually in the distal metaphysis), followed by the humerus and tibia, these 3 locations accounting for >90% of cases [2, 4]. The radius is an extremely unusual site, accounting for only 1.5% of cases [2, 4]. The classical radiographic description is of a lobular, densely ossified mass with a broad, sclerotic cortical base. The ‘string sign’, a thin radiolucency interposed between the tumour and native cortex, is seen in only a minority of cases [6]. Conventional MRI

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Fig. 5 Post-operative MRI of the right distal forearm. (a) Sagittal T2W FSE and (b) axial PDW FSE MR images demonstrate residual intramedullary tumour (black arrows) which has grown since the initial MRI study 4 months previously, as well as soft tissue swelling at the surgical site (white arrowheads)

sequences show low signal intensity similar to cortical bone due to pronounced mineralisation, while regions of soft tissue signal intensity suggest dedifferentiated tumours.

Multiple differential diagnoses exist. Parosteal osteosarcoma may mimic the exophytic ossification of heavily calcified BPOP, an entity which also only rarely arises from the distal forearm long bones [7] and rarely involves the medullary cavity. A history of trauma may suggest florid reactive periostitis, but this is usually associated with prominent soft tissue oedema. The pattern of mineralisation in myositis ossificans is usually the inverse of parosteal osteosarcoma, with peripheral ossification classically described. Parosteal osteoma is a very rare entity, characterised by uniformly dense parosteal bone formation separated from the medullary cavity by cortex which may be normal or thickened [8]. Osteochondroma is usually distinguishable by demonstrating both medullary continuity with the central portion of the lesion and a cartilage cap [9]. Malignancies with

mineralised extra-osseous components such as conventional osteosarcoma and chondrosarcoma should also be considered, given their relative propensity for the long bones [10].

Management options for distal radial surface tumours vary according to the aggressiveness of the lesion. Low-grade surface tumours are typically treated with a ‘shark-bite’ excision which removes the surface tumour and the intramedullary extension, usually with no requirement for reconstruction of the bone. High-grade malignant tumours require wide resection and reconstruction with a vascularised fibular strut graft, possibly with neoadjuvant/adjuvant chemotherapy depending upon the diagnosis and the age of the patient.

The identification of a distal radial surface lesion should prompt careful clinical history and image interrogation since a variety of pathologies can be encountered. In the absence of classical features that convincingly point to one of the more common diagnoses there should be a low threshold for referral to a specialist centre.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

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Fig. 6 Photograph of the cut surface of the second resection specimen showing a smooth, lobulated piece of bony tissue with a pale, homogenous cut surface. The macroscopic features are relatively non-specific

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