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CASE REPORT

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Parosteal osteosarcoma of the ulna: a rare low-grade malignant neoplasm. Case report and review of the literature

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Abstract Parosteal osteosarcoma is a rare low-grade malignant neoplasm. A case localized at the distal ulna is reported, and the tumour's radiological and histological features are described. Wide enbloc resection without postoperative adjuvant chemotherapy was the treatment. Good clinical function without signs of local recurrence was observed at the 3-year follow-up.

Key words Parosteal osteosarcoma • Ulna • Resection

Introduction

Of all long bones, the ulna has the lowest incidence of primary tumour involvement. In most series, primary tumours of the ulna account for less than 1% of all the lesions and less than 2% of all long bone lesions [1]. Myeloma is the most common primary bone tumour of the forearm long bones, followed by osteosarcoma in its different variants: intraosseous (conventional, low grade central, telangectatic, and small cell) and extraosseous (high grade surface osteosarcoma, parosteal and periosteal osteosarcoma), as described by Schajowicz et al. [2]. We describe a rare case in which parosteal osteosarcoma localized in the distal end of the ulna.

Case report

A 42-year-old man presented with a 3-month history of painless swelling in his left forearm in the absence of trauma. The mass had been progressively growing, sometimes decreasing the patient's manual ability. Physical examination revealed a firm, immobile 3.0 x 3.5 cm^2 tender mass involving the dorsal aspect of the distal ulna. There was normal range of motion at the wrist. No motor or sensory deficits were reported. No adenopathy was evident. Laboratory tests were normal.

Radiographs of the wrist demonstrated an inhomogeneous, calcified, lobulated mass with some radiolucent areas arising from the ventromedial part of the distal ulna

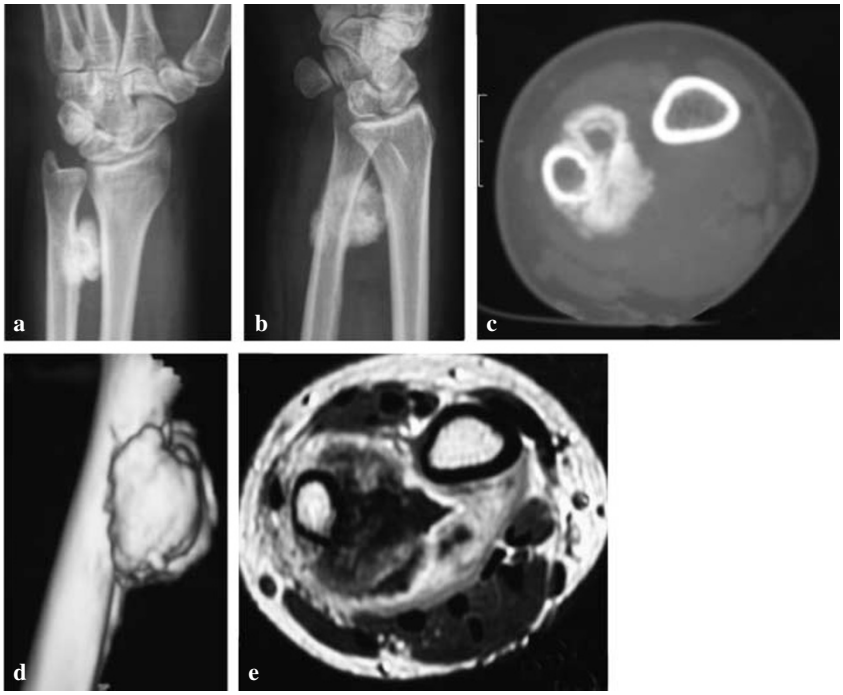

Fig. 1a-e *Diagnostic imaging of a parosteal osteosarcoma of the distal ulna.* **a**, **b** Radiographs in anteroposterior (**a**) and oblique (**b**) projections showed a dense, inhomogenous, calcified mass in the ventromedial part of the distal ulna. **c**, **d** Computed tomograms, without (**c**) and with (**d**) 3D reconstruction revealed a lobulated, calcified mass adjacent to cortical bone without infiltration of bone marrow. **e** MRI demonstrated the absence of neurovascular bundle involvement

(Fig. 1a, b). Computed tomography (CT) with three-dimensional (3D) reconstruction (Fig. 1c, d) revealed that cortical bone was not interrupted. Magnetic resonance imaging (MRI) demonstrated that the bone marrow and the neurovascular bundle of the forearm were not infiltrated (Fig. 1e). Chest CT was normal. The uptake of tracer seen on bone scanning demonstrated that no other bones were involved. The patient underwent open surgical biopsy.

Histological analysis revealed the presence of atypical chondroid cells with a moderate degree of mitotic activity producing osteoid. The combination of clinical features, imaging and histological findings led to the diagnosis of well differentiated parosteal osteosarcoma of the distal ulna. En bloc resection of the distal ulna without reconstruction was planned, and wide margins were obtained (ranging from 3.5 mm to 1 cm in different regions) (Fig. 2). Histological analysis of the tissue in contact with the radial bone revealed the absence of tumour cells infiltrating radial bone. Postoperative positron emission tomography (PET) did not reveal any pathological regions, so adjuvant chemotherapy was not administered.

The patient was followed with PET and MRI for 3 years without signs of local recurrence or pulmonary metastases. At the latest follow-up, the forearm appeared radially deviated by about 15° and the third finger was flexed because of the lesion of the flexor tendon. Extension of the affected wrist was about 60°, compared to 75° on the contralateral wrist (Fig. 3). Grip strength was reduced by 15%, pronosupination was slightly reduced, but supination was quite similar that to on the contralateral side. The patient reported hypoesthesia on the dorsal part of the fifth finger. The patient is able to perform manual activities and has returned to work.

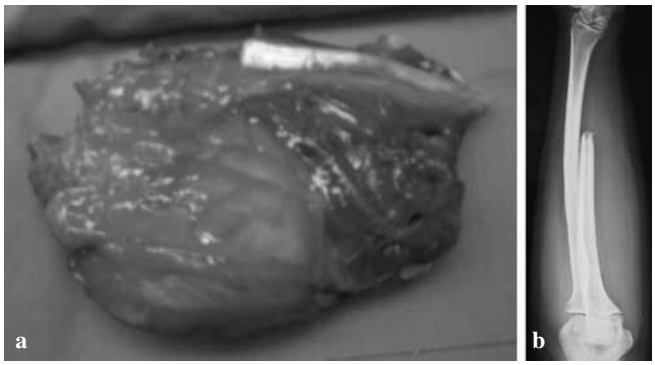

Fig. 2a, b *En bloc resection of distal ulna and surrounding soft tissue with wide margins*. **a** Resected specimen. **b** Radiographic view

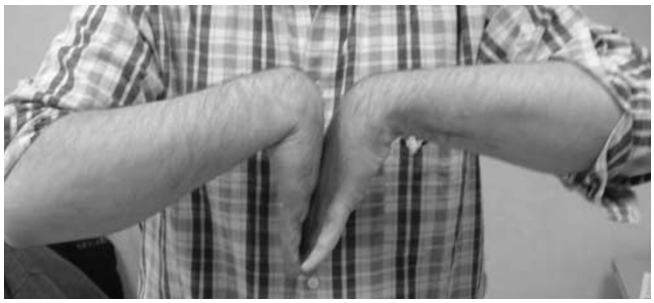

Fig. 3 At the 3-year follow-up, functional results were good and range of motion was only slightly reduced

Discussion

Parosteal osteosarcoma is a malignant bone tumour and is the most common type of surface osteosarcoma. Patients with parosteal osteosarcoma usually have a long history of painless swelling [3]. Patients afflicted by parosteal osteosarcoma usually are in the third or fourth decade of life, older than patients afflicted by conventional osteosarcoma. The lesion most often arises from the metaphysis of the posterior aspect of the distal femur, the proximal tibia and the proximal humerus. Other localizations like the radius and the ulna are rare; presently, only 3 cases of low-grade parosteal osteosarcoma of the distal ulna have been described [4, 5]. The typical radiographic feature is the lobulated, well calcified mass, more dense in the centre than in the periphery, surrounding the host bone [6, 7]. Radiolucent areas may reveal the thickness of the periosteum between tumour and cortex [6]. CT and MRI showed the absence of medullar cavity infiltration. The importance of medullar infiltration is still controversial, but Wold et al. [8] reported a significant correlation between medullary involvement and histological grading: the higher grade, the greater incidence of medullar involvement. The incidence of dedifferentiation of parosteal osteosarcoma is about 20% [8].

Wide en bloc resection (with safety margins of at least 3 mm of normal soft tissue and 2 cm of healthy bone) is the recommended therapy for parosteal osteosarcoma, and provides a 5-year survival rate greater than 90% [3]. Simple excision is inadequate because of a reported recurrence rate of 80%-100%. In the case described by Drinkuth et al. [5], a segment of the distal ulma extending up to 1 cm proximal to the distal radioulnar joint was resected and the resected bone was replaced with iliac bone graft stabilized with internal fixation, which was subsequently removed. In two case series of distal ulna excision for benign and malignant disease, wide resection without reconstruction was recommended, because functional results of the wrist were good and did not correlate with the amount of bone sacrificed [4, 9]. In a cadaveric study, Greenberg et al. [10] demonstrated that resection of the distal ulna leads to stabilization of the forearm (reduction in the radio-ulnar distance) and a reduction of strain in the intraosseous membrane related to the entity of the resection. On this basis, wide excision of the distal ulna may be considered an acceptable salvage procedure in alternative to creation of a one-bone forearm [10]. Our case confirms that wide en bloc resection of the distal ulna without reconstruction leads to a slight difference in function between the afflicted and contralateral wrists and may be considered the treatment of choice for parosteal osteosarcoma in the distal ulna.

Adjuvant chemotherapy is not indicated in low-grade parosteal osteosarcoma, while the recommended treatment for high-grade and dedifferentiated parosteal osteosarcomas is the same as that for conventional osteosarcoma, i.e. neoadjuvant chemotherapy.

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