Extraskkeletal osteosarcoma of the hand

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Introduction

Extraskkeletal osteosarcoma (ESOS) is a rare tumour that does not typically present in the hand. We describe a case of ESOS, a rare soft tissue tumour, arising in the hand as a recurrent soft tissue lump. Establishing a histological diagnosis can be difficult in this rare tumour. This case report highlights the difficulty in the diagnosis and treatment of this rare sarcoma.

Case Report

A 56-year-old man presented with a swelling in his right palm that had increased in size for 3 months. He had full function of his hand but complained of paraesthesia in his middle finger and radial half of his ring finger. There was no history of injury or other comorbidity.

On examination, there was a 3 cm, firm, non-tender, lobulated swelling in the centre of the right palm with no attachment to the skin or flexor tendons. His hand was neurovascularly intact, with full range of movement, and there was no axillary lymphadenopathy.

Magnetic resonance imaging (MRI) showed a 3.9×2.9×2.5 cm mass in the superficial and deep compartments of the palm interposed between the flexor

tendons (Fig. 1). Ultrasound-guided needle core biopsy showed no high-grade malignancy, but was non-diagnostic. He underwent function-preserving marginal excision of the lesion. Histology showed a spindle cell tumour of uncertain malignant potential with few mitoses, chondromyxoid areas and no osteoid (Fig. 2), with clear excision margins.

Four months after excision, there was clinical and MRI evidence of local recurrence. Incisional biopsy showed a highly cellular spindle cell neoplasm with a high mitotic index, nuclear pleomorphism, areas of necrosis and osteochondroid foci. The diagnosis of high-grade ESOS was confirmed by two musculoskeletal pathologists.

No metastases were identified on computed tomography (CT) of the chest, abdomen and pelvis or radioisotope bone scan. He was commenced on doxorubicin and cisplatin chemotherapy. Clinical examination (Fig. 3) and MRI (Fig. 4) after completion of two cycles of chemotherapy showed an increase in the size of the tumour. As the tumour was non-responsive to chemotherapy and not amenable to limb salvage, he underwent radical resection (below-elbow amputation). Resection histology confirmed ESOS with little necrosis. Following surgery, his chemotherapy regimen was changed to ifosfamide and etoposide. However, pulmonary metastases were detected on follow-up chest imaging at 11 months, and he died of advanced metastatic disease.

Discussion

ESOS accounts for 1.2% of soft tissue sarcomas and 3.7–4.6% of osteosarcomas [2]. It is a tumour that arises in the

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Fig. 1 MRI of the original mass in the palm of the hand

soft tissue without bony or periosteal attachment, has a uniform sarcomatous pattern and produces osteoid and cartilaginous matrix [3]. ESOS of the hand is extremely rare with only one previous case report [3]. ESOS typically occurs in the lower extremity, most commonly in the thigh. Unlike skeletal osteosarcoma, which usually presents in the first two decades, ESOS usually develops in the fifth to sixth decades [2, 3]. In the current case, the presence of malignant osteoid in the recurrent lesion but not in the first resection suggests progression of the malignancy to a fully developed osteosarcomatous phenotype rather than dedifferentiation.

Plain radiographs may show soft tissue ossification. MRI shows the extent of the lesion and involvement of surrounding structures. Needle core biopsy may be insufficient for histological analysis, and wide excision may be necessary for formal diagnosis. The pathological differential diagnosis includes myositis ossificans and

Fig. 2 Haematoxylin and eosin-stained section showing a lace-like pattern of pericellular osteoid production by the malignant cells ($\times 250$)

Fig. 3 Clinical appearance of the recurrent ESOS in the patient's right palm

fibro-osseous pseudotumour, both of which are benign, non-aggressive lesions. Staging is by CT and bone scans, and radical excision in the form of amputation may be needed for recurrent/aggressive disease.

The role of adjuvant chemotherapy and radiotherapy after complete resection is unclear. Meta-analysis suggests a small but definite reduction in the risk of local, distant and overall recurrence with adjuvant chemotherapy [4]. Prognosis of ESOS is poor (24–46% 5-year survival) with a median survival for metastatic ESOS of 8 months [1].

Any hand swelling that recurs within a few months following surgery should raise the possibility of a malignant tumour of the hand. Early referral to a tumour specialist/sarcoma centre is essential for establishing the correct diagnosis, appropriate staging and management.

Fig. 4 MRI showing the recurrent ESOS

References

1. Ahmad SA, Patel SR, Ballo MT. Extraosseous osteosarcoma: response to treatment and long-term outcome. *J Clin Oncol*. 2002;20:521–7.
2. Chung EB, Enzinger FM. Extraskkeletal osteosarcoma. *Cancer*. 1987;603:1132–42.
3. Cook PA, Murphy MS, Innis PC, Yu JS. Extraskkeletal osteosarcoma of the hand: a case report. *J Bone Joint Surg Am*. 1998;80:725–9.
4. Pervaiz N, Colterjohn N, Farrokhyar F, Tozer R, Figueredo A, Ghert M. A systematic meta-analysis of randomized controlled trials of adjuvant chemotherapy for localized resectable soft-tissue sarcoma. *Cancer*. 2008;113:573–81.