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CASE REPORT

Soft tissue osteosarcoma : A case report

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Osteosarcomas arising from extraskeletal locations are extremely rare and their true prevalence is unknown. We describe a case of soft tissue-origin-osteosarcoma arising from the vastus medialis muscle in a 58-year old woman. The possibility of a malignant tumour was not suspected initially and she was referred to a specialist unit only one year after the onset of the swelling. She was successfully treated with a surgical resection, chemotherapy and radiotherapy. Because of the rarity of the tumour, a high degree of suspicion is required to make an early diagnosis. The differential diagnosis of a soft tissue mass located in the thigh in a patient over the age of 40 years, should include extraskeletal osteosarcoma.

Keywords : extraskeletal osteosarcoma ; soft tissue osteosarcoma ; malignant tumour ; vastus medialis.

INTRODUCTION

Osteosarcoma is the most common primary neoplasm of bone and is highly malignant. It occurs commonly in the metaphysis of long bones in an adolescent population (*6*). Osteosarcomas originating in soft tissues are extremely rare and their true prevalence is unknown (*1, 2, 12*). In the absence of prospective evidence, which is clearly not feasible because of the extreme rarity of the tumour, formulating a treatment plan is difficult. We describe a case of soft tissue osteosarcoma arising from the vastus medialis in a 58-year old woman. The possibility of a sarcoma was not suspected initially and she was referred to us only one year after the onset of the swelling. She was treated with surgical resection, chemotherapy and radiotherapy.

CASE REPORT

A 58-year old teacher was referred to us with a painless lump in her right thigh, which was gradually growing in size over the last twelve months. She had not sustained any injuries. On examination she had a painless, firm swelling on the medial side of her right distal thigh, measuring 7 cm in diameter. The mass was clinically not fixed to the underlying femur. There was no involvement of regional lymph nodes. Systemic and neurovascular examinations were unremarkable.

Plain radiographs (fig 1) demonstrated a soft tissue mass with calcification. An MRI scan (fig 2) showed a tumour lying within the right vastus medialis muscle in the distal thigh. CT scan of the lungs was normal. There was no regional, hilar or mediastinal lymphadenopathy. An open biopsy was

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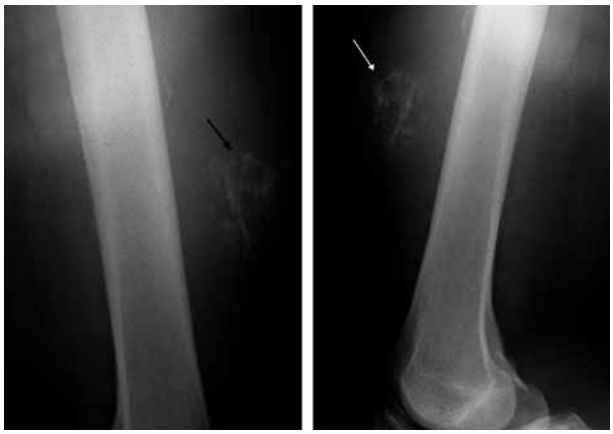

Fig. 1. — Plain radiographs demonstrating soft tissue mass with calcification.

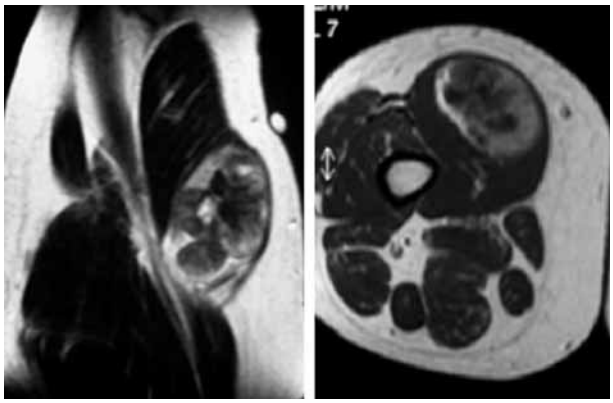

Fig. 2. — An MRI scan showing the tumour lying within the vastus medialis muscle.

reported as a sarcoma with chondroid differentiation. Three days after the open biopsy a radical excision of the tumour was performed. The tumour was resected en bloc with the vastus medialis and a part of the vastus intermedius to achieve a wide margin. The skin defect was covered secondarily with a split skin graft after obtaining histological confirmation of complete and extra capsular resection.

Macroscopically, the resected tumour was lobulated and bony hard at some places and measured 7 cm × 6 cm × 6 cm. Histopathological examination showed (fig 3) spindle cells with large areas of hyalinisation. Focally it showed bone and osteoid formation. The diagnosis of an extraskeletal osteosarcoma was confirmed by immunohistochemistry.

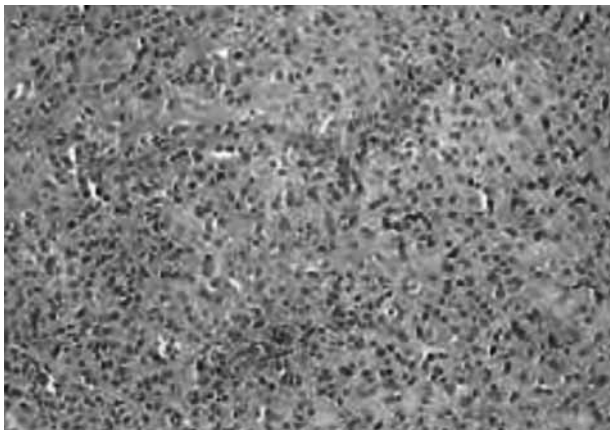

Fig. 3. — Histology of the tumour showing spindle cells with large areas of hyalinisation. Focally it showed bone and osteoid formation.

The patient received adjuvant post-operative chemotherapy (Cisplatin and Adriamycin) and radiotherapy (6000 Gy, in 30 fractions over a period of 47 days). She developed febrile neutropenia during the course of chemotherapy. At 48 months of follow-up there was no evidence of local or distant disease clinically and radiologically. She now works part time in her previous job.

DISCUSSION

Extraskeletal osteosarcoma was first described by Wilson in 1941 (*13*). Unlike primary osteogenic sarcoma, which commonly affects the individuals younger than 25 years of age, extraskeletal osteosarcoma usually develops during the fifth and sixth decade of life (*1-4, 12*). Even though the five-year survival for osteogenic sarcoma improved to 60% after the introduction of adjuvant chemotherapy, the reported five-year survival for extraskeletal osteosarcomas remains at 25-35% with similar treatments (*2, 4, 8, 12*). This fact raises the possibility that skeletal and extraskeletal osteosarcomas could be different entities biologically and therefore behave in different patterns. The higher mortality may also be because of occurrence of this tumour in an older population compared to the osteogenic sarcomas.

Soft tissue osteosarcoma most commonly presents as a progressively growing soft tissue swelling (*1, 2, 3, 12*). History of preceding trivial trauma is present in a significant number of patients (*9*), but the clinical and pathological significance of the trauma is unknown. Evolution of extraskeletal osteosarcomas from myositis ossificans, dermatomyositis with calcinosis and heterotrophic bone have been reported (*5, 7-9*). In patients aged over 40 years with a soft tissue mass in the thigh differential diagnosis includes malignant fibrous histiocytoma, chondrosarcoma of soft tissues and myositis ossificans (*5, 7-9*). To diagnose an extraskeletal osteosarcoma the tumour must arise in the soft tissues without any attachment to periosteum or bone (*9, 12, 13*). Histologically the tumour should have a uniform sarcomatous pattern containing osteoid or cartilaginous matrix (*9, 12, 13*). Five subtypes (osteoblastic, chondroid, fibroblastic, telangiectatic and small cell types) of soft tissue osteosarcoma have been described depending on the type of the matrix (*9, 10, 12, 13*). The size of the primary tumour appears to give prognostic significance (*2, 11*), although not every study supports this observation (*5*). Survival outcome is significantly better in patients with primary lesions less than 5 cm (*2*).

The patient in the present study had an intracompartmental tumour without any evidence of regional or distant spread. However the tumour size was bigger than 5 cm in all dimensions, which required a large resection and closure of the defect by a skin graft. Even though adequate time was given for the soft tissues to heal before the commencement of radiotherapy she developed wet desquamation reaction which healed completely once the radiotherapy was completed.

Radical resection is effective for local control and has the best chance of cure (*9*). However, radical resection had no impact on distant metastasis. Hence systemic chemotherapy is commonly advocated in treatment of these tumours. Because of the rarity of the soft tissue osteosarcomas, no prospective evidence is available regarding the usefulness of the adjuvant chemotherapy or radiotherapy. Recurrence rates of more than 50% have been reported usually within 3 years (*1, 2, 9, 12*).

In this paper a rare case of soft tissue osteosarcoma arising from the vastus medialis muscle is reported. In a patient over 40 years of age with a soft tissue mass in the thigh, the differential diagnosis of the mass should include extraskeletal osteosarcoma. Because of the rarity of the tumour, a high degree of suspicion is required to make an early diagnosis.

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