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Extra-abdominal desmoid tumour of the leg

Agrawal P S, Jagtap S M, Mitra S R

ABSTRACT

Extra-abdominal desmoid tumour is a rare tumour and only a few cases occurring in the limbs have been reported. A 35-year-old woman presented with gradually increasing swelling in the upper leg. She had a mild, dull, aching pain in the tumour. Wide local excision was done and the tumour was found mainly in the subcutaneous tissue, which histopathologically proved to be an extra-abdominal desmoid tumour. This case had an abnormal radiological appearance of peripheral calcification of tumour and saucer-shaped lesion in the underlying tibial cortex. The patient had no recurrence at two years follow-up.

Keywords: desmoid tumour, extra-abdominal desmoid tumour, musculoskeletal tumour

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INTRODUCTION

Extra-abdominal desmoid tumour is a non-metastasising fibrous tumour. It is characterised by infiltrative invasion of soft tissues and a high propensity for local recurrence after surgical excision.[1,2] We present a rare case of this tumour found in the upper leg of a woman.

CASE REPORT

A 35-year-old woman presented with a one and a half year history of a slowly-growing swelling over the left upper medial surface of the tibia. The swelling was associated with a dull, aching pain. She had no other complaints. Examination revealed an irregular tumour of 12 cm x 10 cm, situated over the medial subcutaneous surface of upper tibia, just below the medial joint line of tibia and covering the medial border of patellar tendon. Clinically, the tumour was hard, immobile, attached to the bone but free from the overlying skin. The skin over the swelling was normal. There were no similar swellings in any other parts of the body. Local lymph nodes were normal.

Radiographs of the leg showed a soft tissue swelling over the upper end of the tibia and peripheral irregular coarse calcification without any underlying bone pathology (Fig. 1). Computed tomography (CT) of the leg revealed a well-defined irregular soft tissue density on the anteromedial aspect of the proximal end of the left tibia with multiple thick dense calcific foci in the periphery. There was evidence of a small area of cortical erosion of the anteromedial tibia (Fig. 2). Fine-needle aspiration cytology from the swelling was inconclusive as no aspirate could be obtained.

Excision biopsy of the tumour was planned. Under spinal anaesthesia, with the patient in a supine position, an incision along the long axis of tumour anteromedially over the leg was made. The tumour was firm to hard, poorly-encapsulated and was mainly located in the subcutaneous tissue. It was around 10 cm x 10 cm in size, and attached to the periosteum of medial tibia over a small area. A small underlying 0.5 cm x 1cm cortical erosion was also present just medial to the tibial tuberosity, but the bone was not obviously involved. The tumour at one place was involving the dermis, so that part of skin was also excised. En bloc excision of the tumour was carried out and sent for histopathological examination. The wound was closed in layers over negative suction drainage. Postoperative recovery was uneventful and stitches were removed after ten days. The histopathological examination showed features of extra-abdominal periosteal fibromatosis (desmoid) with calcification. The patient was followed-up for two years, during which there was no evidence of tumour recurrence.

DISCUSSION

Extra-abdominal desmoid tumour is also known as extra-abdominal fibromatosis, desmoid fibromatosis, well differentiated non-metastatising fibrosarcoma, musculoaponeurotic fibromatosis and even grade I fibrosarcoma.[1,3,4] Extra-abdominal desmoid tumour is a fibroblastic tumour arising from the connective tissues of muscles, fascia, aponeurosis or periosteum. It is a locally-aggressive tumour, and well known to invade nearby structures, such as muscles, subcutaneous tissue and neurovascular structures. The tumour is not known to metastasise, but has a high recurrence rate after surgical excision. These tumours have a slight predilection for the male gender and can occur at any age, but are more commonly occuring in young adults. These extra-abdominal

Department of Orthopaedics, Government Medical College Hospital, Medical Square, Nagpur, Maharashtra 440003, India

Agrawal PS, MBBS, MS
Lecturer

Jagtap SM, MBBS, MS
Lecturer

Mitra SR, MBBS, MS
Professor and Head

Correspondence to:
Dr Pravin S Agrawal,
Kanchan Yash,
West Park Road,
Dhantoli,
Nagpur 440012,
India
Tel: (91) 712 243 7095
Fax: (91) 712 243 1111
Email: drpravin_ngp@yahoo.com

tumours commonly arise in the musculature of the shoulder, chest wall, upper arm, thigh and head-neck.[1,3-5] Fibromatosis arising around the knee is very rare.[1,3-6]

Abramowitz et al defined two patterns of bone involvement in these tumours arising from soft tissues. In the first pattern, there is erosion of the contiguous cortex, producing a lytic, saucer-like cortical defect with sclerotic margins. The second was a distinct "frond-like" periosteal reaction, which consists of spicules of bone radiating into the soft tissue mass.[6] In our patient, we found saucer-shaped cortical erosion of the tibial cortex. Calcification was seen, but it was not typically "frond-like", and was present more at the periphery of the tumour. Thus, both types of lesions described by Abramowitz et al were found in a single case which is very rare.[6] Also, the pattern of calcification was very much different from what is classically described.

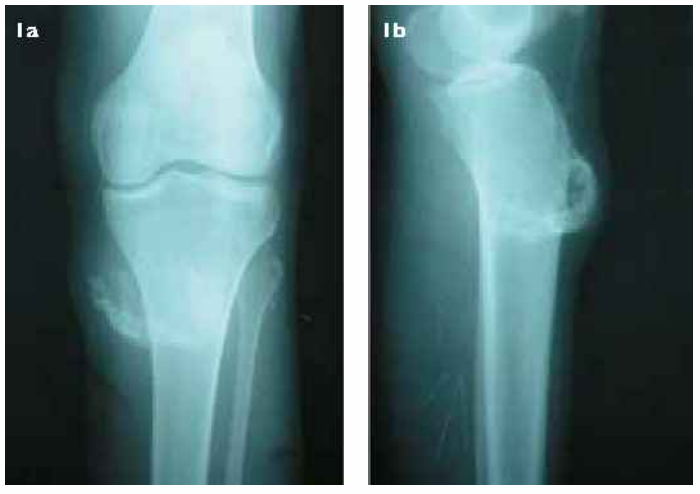

Fig. 1 (a) Anteroposterior and (b) lateral radiographs show a soft tissue mass with peripheral calcification.

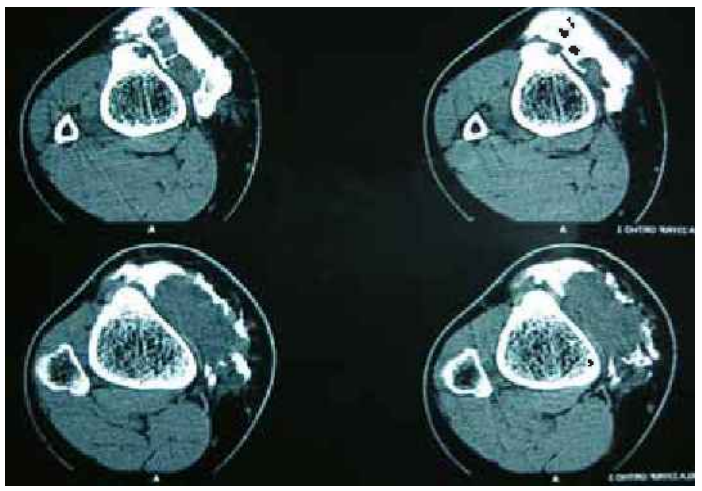

Fig. 2 Axial CT images show peripheral calcification of the tumour with a saucer-shaped osteolytic lesion of the adjacent tibial cortex.

In our case, the tumour was not arising from any muscle compartment of the leg and it was mainly situated in the subcutaneous tissue, which also makes it a unique case. Complete excision of tumour was possible and the patient has no recurrence at two years of follow-up. These tumours appear histologically benign. The tumour infiltrates the surrounding tissues without forming a pseudocapsule.[1,3] Pathogenesis is unknown, but trauma, endocrine and genetic causes are associated factors. These tumours may arise as a part of Garden's syndrome.[3,6] Wide surgical excision is the surgical goal of treatment. Low recurrence rate is obtained in patients who had wide surgical margins. Postoperative radiotherapy is recommended when wide surgical margins are not achieved. Recurrent lesions can be managed on the same surgical principles if they are progressive or can be observed if lesions are stable.[2,5,7,8]

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