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A Case Report on Aggressive Fibromatosis with Bone Involvement

Y Y Tan, C K Low, P Y Chong

ABSTRACT

Aggressive fibromatosis is a locally infiltrative fibroblastic tumour that arises from fascial planes of soft tissue but does not metastasize. It is known to invade muscle, subcutaneous tissue and neurovascular structures. However, bone involvement is very rare and there has been few reports of bone involvement[1-5]. We present a case of a young man with aggressive fibromatosis of the right lower leg with fibula involvement.

Keywords: extra-abdominal desmoid tumour, radiological changes

CASE REPORT

A 32-year-old Chinese man presented with a one-year history of swelling in his right leg associated with nocturnal pain. Clinical examination showed a 7 cm by 6 cm hard swelling over the lateral aspect of the right leg which was fixed to the underlying peroneal muscles but not to the overlying skin.

Plain radiographs of the right leg (Fig 1) showed an expansion of the lower third of the fibula associated with posterior bowing and cortical thickening. There was excavation of the anterior aspect of the fibula cortex together with a moth-eaten appearance of the bone. Magnetic resonance imaging (MRI) showed a soft tissue mass lodged within the peroneus brevis muscle. This mass abutted on the lateral aspect of the lower fibula which was deformed with thickening of the cortex, scalloping and spiculations. There was intense enhancement of the mass with contrast.

A 7 cm by 6 cm firm well-defined tumour arising from within the peroneus brevis muscle was found during surgery. Wide excision of the tumour was performed and the adjacent outer cortex of the fibula was resected en bloc with the tumour. Pathologic examination of the specimen showed a whitish tumour mass 5 cm by 3.5 cm within the soft tissue. The tumour was on the surface of the bone and 0.9 cm from the skin. Histologic examination showed an extra-abdominal fibromatosis composed of a proliferation of fibroblastic cells. No mitotic activity or necrosis was noted. Tumour tissue was found within the scalloping area of the bone and separated from the medullary canal by the cortical bone. The resection margins were free of tumour.

Ten months later, the patient presented with a 5 cm by 5 cm firm, ill-defined swelling over the right lower leg. Radiographs showed a soft tissue mass with a moth-eaten appearance of the adjacent fibula (Fig 2). MRI studies reported a large soft tissue tumour in the lower leg invading surrounding structures including the medullary cavity of the mid-shaft of the fibula.

Wide excision of the tumour was performed together with the segment of the involved fibula. Pathologic examination confirmed recurrence of aggressive fibromatosis. The tumour measured 19 cm by 8 cm by 7 cm and was seen surrounding the fibula. Medullary involvement was demonstrated.

Post-operative recovery was uneventful and the patient is well after a follow-up of 2 years. He is walking without support and there is no evidence of tumour recurrence.

DISCUSSION

Aggressive fibromatosis is also known as extra-abdominal fibromatosis, extra-abdominal desmoid tumour, desmoid tumour, well-differentiated non-metastasising fibrosarcoma, desmoid fibromatosis, and even fibrosarcoma grade I desmoid type. It should be distinguished from desmoid tumour of bone (desmoplastic fibroma) which is an intraosseous fibrous tumour which originates from bone.

Aggressive fibromatosis is a fibroblastic tumour, arising from the connective tissue of muscle and the overlying fascia or aponeurosis. It is locally aggressive, growing to large sizes and well known to invade nearby structures such as muscle, subcutaneous tissue and neurovascular structures. Despite its outwardly aggressive nature, histologically it consists of uniform spindle cells with a variable amount of collagen, and no nuclear atypia or abnormal mitoses. The tumour is not known to metastasize, but considerable morbidity and dysfunction may be present when surrounding structures are involved. There is a high rate of recurrence after surgical excision, as the tumour margins often extend beyond that palpable, and its infiltration into deep seated areas makes it difficult to achieve wide excision without sacrificing vital structures.

Whilst aggressive fibromatosis is known to be locally infiltrative, there has only been a handful of cases reported whereby bone involvement on plain radiography was documented[1-5].

Department of Orthopaedic Surgery
Tan Tock Seng Hospital
Moulmein Road
Singapore 308433

Y Y Tan, MBBS
Medical Officer

C K Low, FAMS, FRCS, MMed (Surg)
Consultant

Department of Pathology
Singapore General Hospital
Outram Road
Singapore 169608

P Y Chong, FAMS, FRCPA
Consultant

Correspondence to:
Dr C K Low

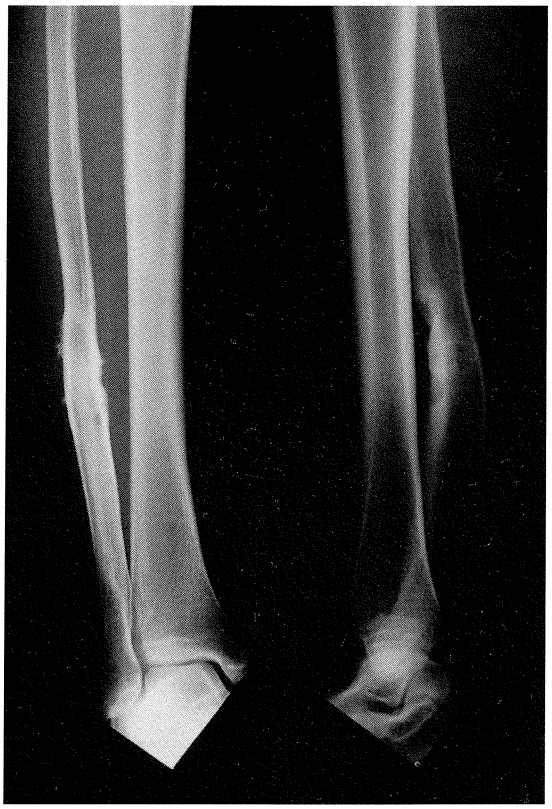

Fig 1 – Plain radiograph of the right leg showing expansion of the lower fibula with posterior bowing, cortical thickening of the interior cortex and a moth-eaten appearance.

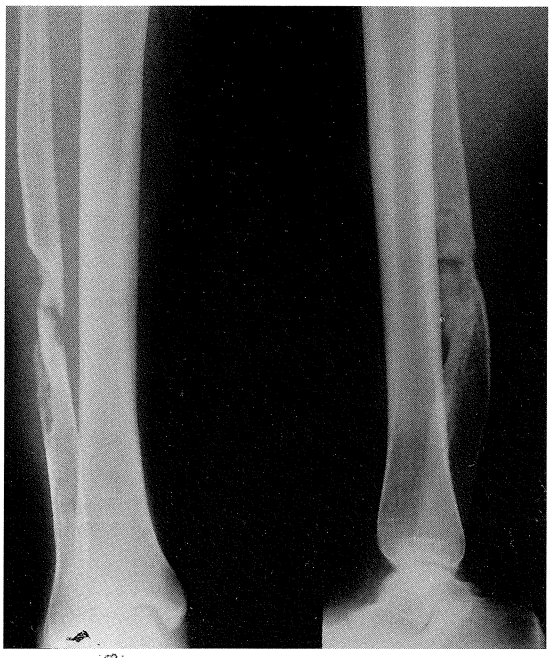

Fig 2 – Recurrence of the tumour with a lytic lesion of the fibula.

Rosen and Kimball[1] first described localised periosteal thickening and frank bony destruction by aggressive fibromatosis. In 1979, McDougall and McGarrity[2] reported two cases of aggressive fibromatosis involving the bone. One was a 25-year-old lady who was diagnosed to have aggressive fibromatosis of the forefoot, with radiographic changes of the metatarsals. The patient subsequently went on to develop multicentric desmoid tumours over the abdominal wall and ipsilateral thigh. The second case was that of a 16-year-old girl with aggressive fibromatosis over the inner aspect of her right elbow. Radiology revealed erosion of the humerus at the site of the tumour. After excision of the tumour, she returned 2 years later with a local recurrence of the tumour. Radiography revealed increased erosion of the adjacent humerus. This was treated conservatively. The patient subsequently returned with a similar tumour of the ipsilateral deltoid region.

Abramowitz et al in 1983[3] clearly defined two patterns of bone abnormalities present in aggressive fibromatosis in eight of their patients. The first was non-specific pressure erosion of the adjacent cortex, presenting as a lytic, saucer-like cortical defect with a sclerotic margin. The second was a distinct 'frondlike' appearance of the adjacent bone, seen as spicules of bone radiating into the soft tissue mass. This is thought to be produced by periosteal stimulation of the bone by the tumour. In none of his cases was there evidence of intramedullary involvement.

Our patient is of interest because of the radiological changes of cortical thickening, moth-eaten appearance and posterior bowing of the fibula. Excavation of the bone causing the cortical defect seen in our patient is likely to be due to pressure changes as described previously. Cortical thickening and posterior bowing may be secondary to reactive hypertrophy of the bone. There was a definite moth-eaten appearance of the fibula. Tumour tissue was found within these scalloping area and separated from the medullary canal by the cortical bone. The resection margins from the first surgery were free of tumour. There was a recurrence of the tumour 10 months later, and the second specimen showed multiple tumour nodules along the subcutaneous tissue apart from the main tumour. The tumour was also seen to involve the medullary canal of the fibula.

The histology of the first specimen showed that the cortex was preventing the tumour from spreading into the medullary canal of the bone. Scalloping of the bone was due to the pressure effect. Examination of second specimen showed that recurrence of tumour in the soft tissue and the medullary canal. As the outer cortex of the fibula had been removed during first resection, tumour tissue could access into the medullary canal. This case illustrates that radiological changes are due to pressure effect rather than direct infiltration. The cortical bone can resist invasion of the tumour. The tumour can spread into the medullary canal if the cortex is breached following surgical removal or fracture.

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