CASE REPORT

Aggressive Fibromatosis: A Case of Bone Involvement

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Abstract

Bone involvement in aggressive fibromatosis is very rare. We present such a case involving sacrum and sacroiliac joint.

Key words

Fibromatosis, Sacrum, Sacroiliac joint

Introduction

Aggressive fibromatosis is a locally infilterative tumour that originates from facial planes of soft tissues but does not metastasize. It is known to involve muscle, subcutaneous tissue and neurovascular structures. However, bone involvement is very rare and a case report-involving fibula has been reported recently (1). We present a case of a young female with aggressive fibromatosis of gluteal region involving sacrum and left sacroiliac joint.

Case Report

A young married female aged 35 years presented with history of progressive swelling in left gluteal region and occasional dull ache for last six months. There was no history of trauma or other swelling anywhere else. Examination revealed a firm swelling about 6 x 4 inches involving left gluteal region. Overlying skin was free and superficial veins were sparsely engorged. The swelling was non-tender and ranges of motion of both hips were normal. Lumbosacral spine was normal and neurological examination was within normal limits. X-ray pelvis was normal except for infiltrative erosion of left side of sacrum near left sacro-iliac joint (Fig 1). Complete blood count and ESR were normal. On operation, we found a huge swelling involving gluteal muscles on left side and infiltrating into sacrum and left sacroiliac joint.. The tumour

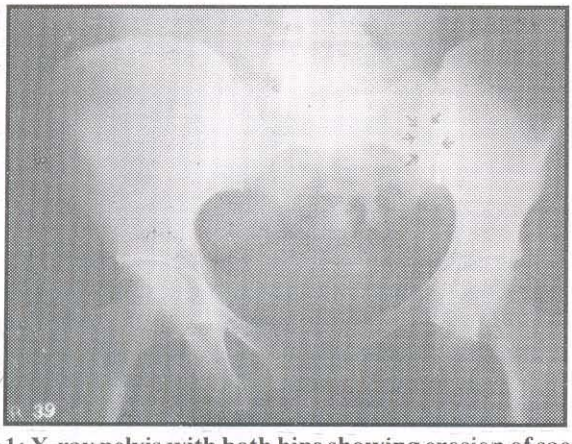

Fig 1: X-ray pelvis with both hips showing erosion of sacrum near left sacroiliac joint.

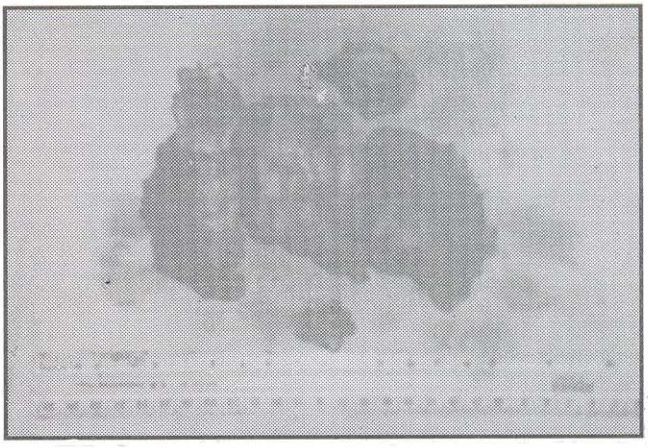

Fig 2: Gross appearance of the tumour after removal

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was removed with surrounding normal tissue of more than 3 cms and underlying bone decorticated and wound closed in layers (Fig 2). Biopsy revealed aggressive fibromatosis (Fig 3,4). The patient was followed for 2 years and showed no recurrence till date.

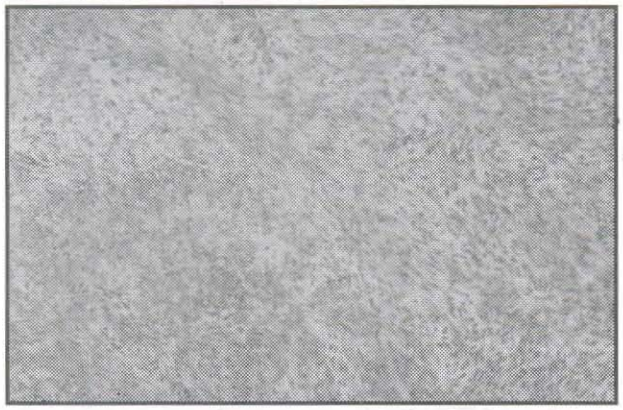

Fig 3. A low power photomicrograph showing interdigitating bundles of fibroblasts with abundant intercellular collagen matrix (H&E).

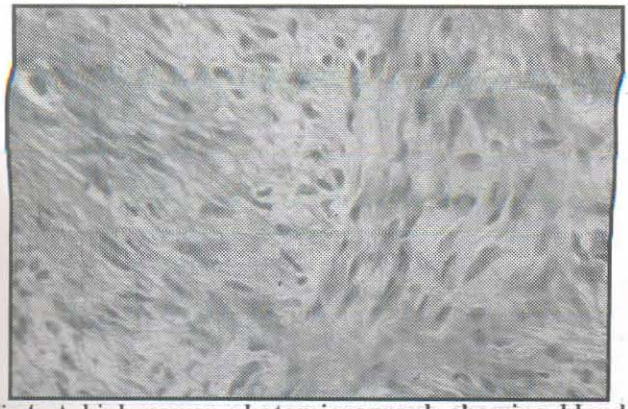

Fig 4. A high power photomicrograph showing bland appearance of fibroblasts (H&E).

Discussion

Deep seated musculo-aponeurotic fibromatosis, also known as aggressive fibromatosis or desmoid tumour, is a relatively rare lesion, representing less than 3% of all soft tissue tumours with a reported annual incidence of 0.2-0.5 / 100000 population (2-4).

Histologically, the lesion belongs to a group of fibroblastic proliferations arising from fascial planes and musculo-aponeurotic structures. Despite its relatively bland cytological features and low mitotic activity the tumour is poorly circumscribed with a strong infiltrative growth pattern along tissue planes and invasion of adjacent tissues (5,6).

Mackenzie has defined the term fibromatosis, as an infiltrating fibroblastic proliferation without any feature of unequivocal neoplasia. The prefix "aggressive" is preferred because it gives better impression of tumour biology with its extra-ordinary local morbid behavior. Confusion still remains, and some consider the aggressive form to be a low-grade fibrosarcoma (4,7). The real incidence remains questionable because pathologists sometimes have difficulty distinguishing these from fibrosarcomas (6,8). None of the predisposing factors like trauma, an association with previous pregnancy and surgical scar (9) were found in our case.

At present wide local excision with a cuff of normal tissue seems to be the most effective treatment. A margin of 2-3 cms beyond the palpable tumour is usually advocated (7). The recurrence rate is very high in these *tumours but the present case showed no recurrence till 2* years of follow up.

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