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Aggressive Infantile Fibromatosis

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INTRODUCTION

The musculoskeletal fibromatosis comprises a wide range of lesions with a common histological appearance. They can be divided into two major groups: superficial (fascial) and deep (musculo-aponeurotic). The superficial fibromatosis are typically small, slow-growing lesions and include palmar fibromatosis, plantar fibromatosis, juvenile aponeurotic fibroma and infantile digital fibroma. The deep fibromatosis are commonly large, may grow rapidly and are more aggressive. They include infantile myofibromatosis, fibromatosis colli, extra-abdominal desmoid tumor, and aggressive infantile fibromatosis.

CASE REPORT

A four-year old girl presented with history of left gluteal swelling for three years. This swelling started as a small bulge, which the parents initially thought secondary to vaccination. However, it was found progressively increasing in size. There was no pain or any difficulty in walking. There is no fever or other generalized symptoms.

Examination showed a large lobulated firm mass in the left gluteal region. There was apparent fixity to underlying muscles.

All routine laboratory examinations were within normal limits. Serum electrolytes were within normal range.

Plain radiography of pelvis shows soft tissue mass in the left gluteal region. No calcification was seen. The left ilium was appearing dysmorphic.

The CT scan showed a large lobulated swelling in the left gluteal region, with no calcification (figure 2). The swelling was just projecting to greater sciatic foramen but not extending to pelvis. The underlying muscles were infiltrated.

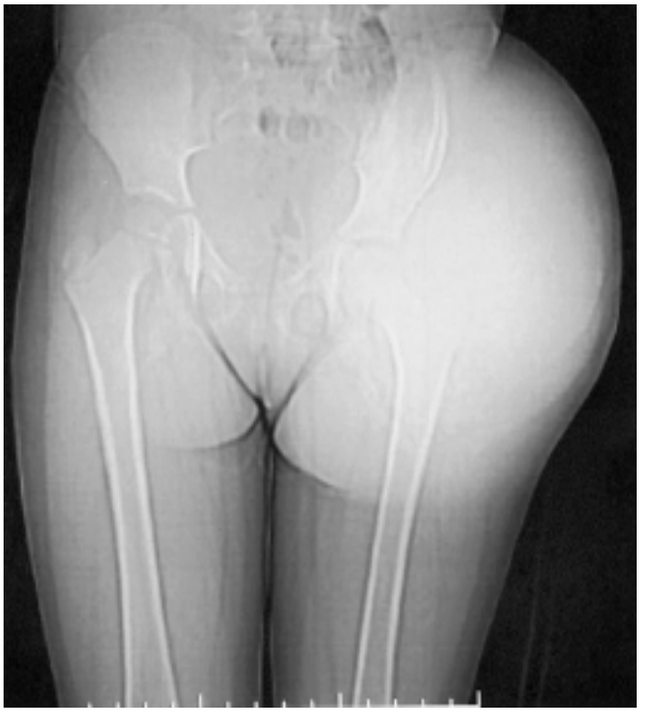

Fig 1: Plain radiograph of the pelvis showing soft tissue mass in the left gluteal region with a dysmorphic ileum

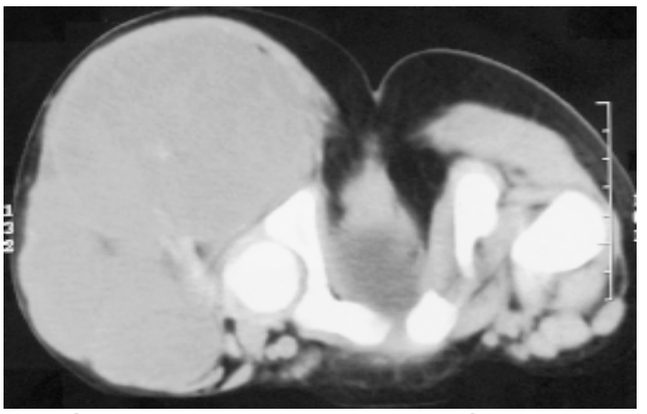

Fig 2: CT scan showing a large lobulated left gluteal region mass lesion with no calcification

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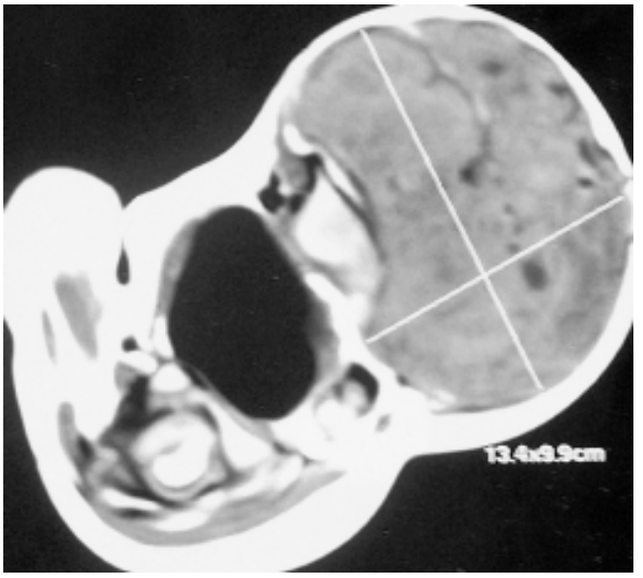

Fig 3: T2W MRI showing an intermediate signal intensity mass lesion in left gluteal region

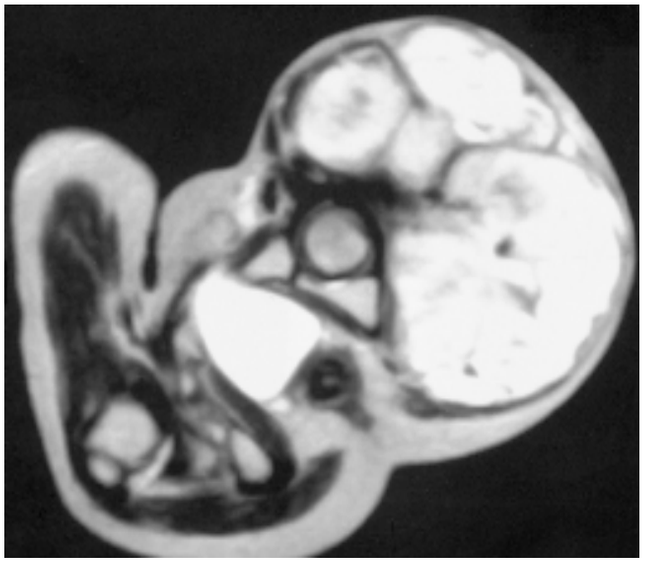

Fig 4: T2W MRI showing a hypointense mass lesion in the left gluteal region involving the gluteal muscles

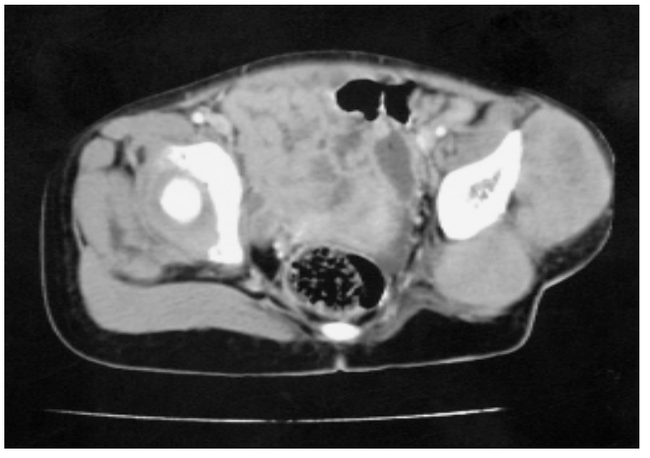

Fig 5

MRI shows heterogeneous intermediate signal intensity on short TR images with hypointense areas. On long TR images the lesion is hyperintense. The gluteal muscles were infiltrated.

The histopathology shows cellular lesion with spindle cells and less fibrosis suggestive of aggressive fibromatosis. The margins were not well defined.

The lesion was surgically removed and the patient was all right for the next one year after which the swelling was again apparent. The lesion was slowly increasing in its size and the child was referred for a repeat CT scan, to know the extent of lesion before surgery.

DISCUSSION

Aggressive infantile fibromatosis appears as painless soft tissue swelling or masses in the extremities usually during first two years of life [1]. They are slightly more prominent in boys. The tumor rarely metastasise [2]

The lesion typically manifests as a firm nodular mass usually within the skeletal muscle, adjacent fascia or periostium. The most common locations are in the head and neck, often with involvement of the tongue, mandible and mastoid process. Other sites frequently affected include the shoulder thigh and foot [3].

These tumors are locally aggressive, infiltrating to muscles, vessels, nerves, fascia, tendons and subcutaneous fat.

Microscopically the level of mitotic activity varies greatly among the interlacing bundles of fusiform and spindle shaped cells, and reticulin and collagen fibres. The histologic features make differentiation from fibro sarcoma difficult. The lesions tend to recur after surgery.

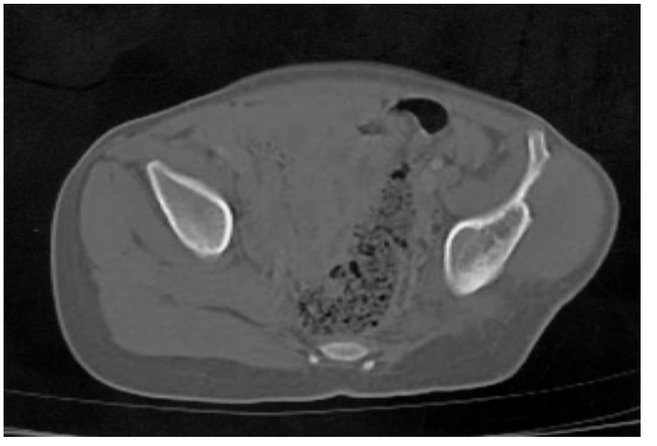

Fig 6

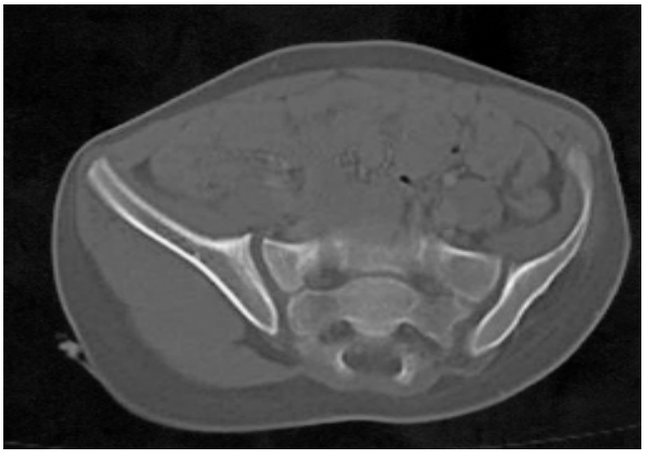

Fig 7

The radiograph demonstrates a soft tissue mass, or swelling with occasional bone defect or scalloped surface. The CT scans are usually non-specific. The lesions may be hypo attenuating relative to skeletal muscles but are typically iso-attenuating or even hyper attenuating [4]. Lesions usually show enhancement with intravenous administration of iodinated contrast, the enhancement is some times marked. Due to infiltrative growth pattern and attenuation similar to skeletal muscle, the margins of the lesion are often indistinct at CT. Pressure erosion of bone can occur (figure 6).

MRI is the best imaging modality. Infantile fibromatosis is an inter-muscular lesion, with frequent muscular invasion. Linear extension along fascial planes is frequent. The initial reports suggest decreased signal intensity in T1 and T2 weighted images. Hypocellularity and abundant collagen produces low signal on T2 weighted images according to Sundaram et al [5]. Marked cellularity shows high signal on T2 weighted images. The most common patterns are of heterogeneous with intermediate signal intensity in standard pulse sequences (equal skeletal muscle on T1 and fat on T2). The heterogeneous pattern shows varying degrees of cellular tissues, myxoid tissue and collagen. Low signal intensity areas suggest areas of dense collagen.[6].

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