

AGGRESSIVE FIBROMATOSIS OF THE EXTREMITIES

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ABSTRACT

Three cases of aggressive fibromatosis of the lower extremities were presented. The patients were female, age ranged from 13 to 27 years old. Ultrasonography showed a mass with infiltrative border and inhomogeneous low echoic pattern. Non contrast CT scan showed isodensity to the muscle. MRI study showed mixed signal on both T1WI and T2WI and strong Gd-DTPA enhancement. Angiography revealed faint tumor stain.

INTRODUCTION

Desmoids or aggressive fibromatosis biologically lie in the interface between exuberant fibroproliferations and low-grade fibrosarcomas. On the one hand, they present frequently as large, infiltrative masses that may recur incomplete excision but, on the other hand, may be small masses composed of well-differentiated fibroblasts that do not metastasize. They may occur at any age but are frequent in the second to fourth decades. Desmoids are divided into extra-abdominal, abdominal and intra-abdominal, but all have essentially similar gross and microscopic features. Extra-abdominal desmoids occur in men and women with equal frequency and arise principally in the musculature of the shoulder, chest wall, back and thigh (1,2). In addition to their possibly being disfiguring or disabling, desmoids are occasionally painful. Although curable by adequate excision, they stubbornly recur in the local site when incompletely removed. The rare reports of metastasis of a desmoid must be interpreted as misidentification of a low-grade fibrosarcoma.

CASE REPORTS

CASE 1

A 13-year-old female patient, had a lemon-sized mass at right popliteal fossa. There was no tenderness at the mass. The mass was firm and fixed to the surrounding tissue. She was operated for this problem two years ago. Ultrasonography showed an inhomogeneous low echoic nodule, with infiltrative border at the muscle plane (Fig. 1). At operation, the infiltrative soft tissue mass, size 5 cm, rubbery in consistency, was noted at around popliteal vessels and nerve. The operation was unsuccessful due to vascular injury. Histology revealed aggressive fibromatosis.

CASE 2

A 27-year-old woman, had a recurrent mass at right popliteal fossa for 5 months. She was operated for aggressive fibromatosis at the same area 8 months ago. At physical examination, a 11 cm-diameter mass was seen at right popliteal fossa. The mass was hard and irregular border. Non i.v. enhanced CT scan of the mass showed an isodensity mass, at right popliteal

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area, involving the subcutaneous fat plane and muscular plane. The density of the mass was 30 H.U. There was no bony involvement (Fig. 2). At angiography, the two lobulated masses were noted with vascular staining. Small neovascularity was observed (Fig. 2).

CASE 3

A 20-year old woman, had a palpable mass at posterior aspect of the left thigh for 2 years, without tenderness. Plain films at the mass showed no calcification. At angiography, the mass had the tumor stain and small faint neovascularity was observed. There was no bony involvement. At MRI study, the mass had well defined border and mixed signal pattern. On T1WI, two signal pattern was observed, dark grey, grey and faintly bright areas were noted. On T2WI, similar mixed signal pattern was noted, except that the dark grey area turned grey and faintly bright area turned brighter signal-area.

Some residual dark grey area was noted. The lesion was totally in the muscle plane (Fig. 3). Gd-DTPA enhancement showed strongly enhanced pattern (fig. 4). Biopsy revealed aggressive fibromatosis.

DISCUSSION

Morphologically, the lesions of aggressive fibromatosis occur as unicentric grey-white, firm, poorly demarcated masses varying from 1 to 15 cm in greatest diameter. They are rubbery and tough and infiltrate surrounding structures. Histologically, they are composed of plump fibroblasts having minimal variation in cell and nuclear size interspersed within a densely collagenous background. Mitoses are infrequent. Regenerative muscle cells when trapped within these lesions may take on the appearance of multinucleated giant cells.

Aggressive fibromatosis at the angle of mandible was demonstrated by ultrasonography by Lewis (3) to be a hypoechoic mass with bone destruction.

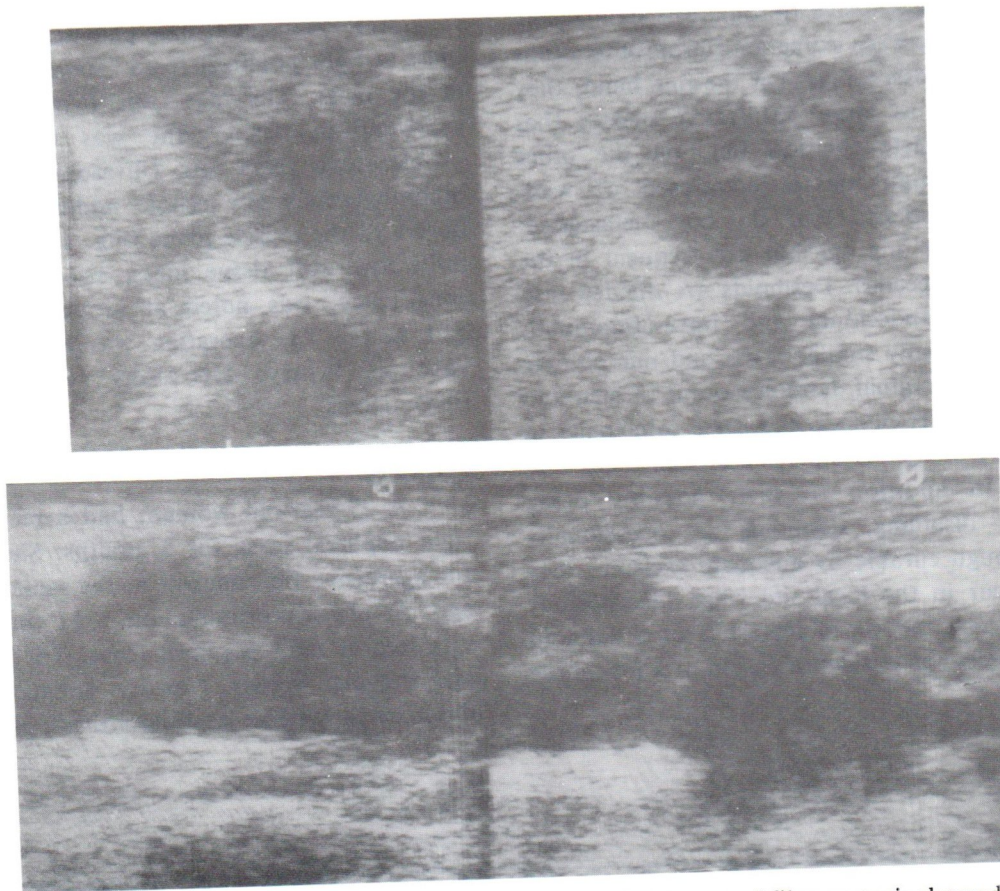


Fig. 1. Case 1. L-and X-section ultrasonography of the mass of fibromatosis showed mixed iso and low echoic lobulated border mass in the muscle plane.

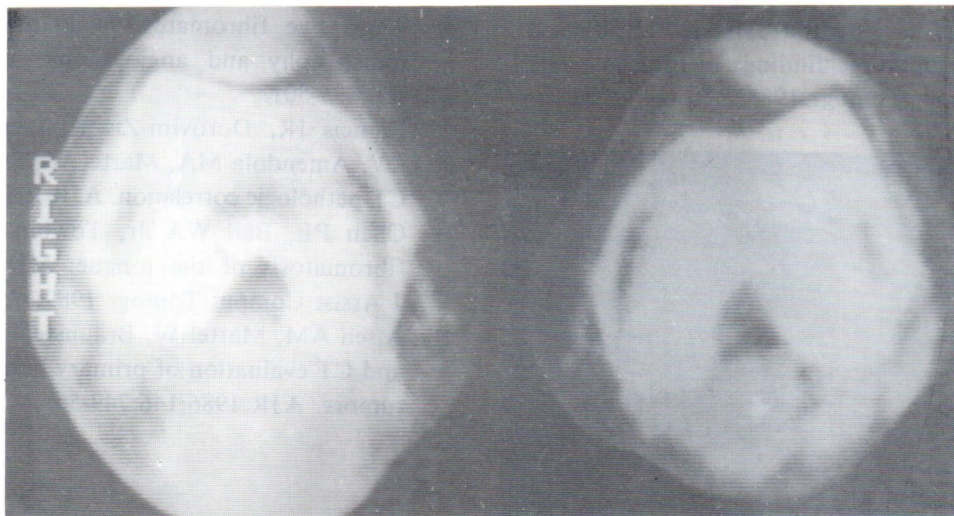


Fig. 2A. Case 2. Non contrast CT scan of the right popliteal area showed a soft tissue mass in the subcutaneous fat plane and muscle plane with the CT density of 30 H.U. which is equal to the muscle of the normal side.

Leibman (4) reported sonographic features of this condition in the breast in two patients. It showed an irregularly shaped hypoechoic mass with lack of posterior attenuation in one case and a hypoechoic mass with internal echoes in the center with decreased through-transmission laterally in another case. The lesion was seen as a hypoechoic nodule of uniform consistency localized superficial to the medial slip of the plantar fascia by Reed (5).

The CT appearance of fibromatoses in childhood was reported by Campbell (6), and in all three cases, the lesions were hypodense on post contrast scans. Hudson (7) reported the CT and angiographic appearance of fibromatoses in 13 cases. They concluded that the tumors were usually better demonstrated after contrast infusion and that no relationship could be established between the vascularity of the tumors and their histologic features. Francis (8) reported four patients with fibromatoses by CT scan. Three of the patients had the lesions which were hyperdense relative to the skeletal muscle on non contrast scans and one patient had the lesion which was hypodense. The lesion post contrast enhancement showed a variable appearance; ranged from minimal contrast enhancement, mixed hyperdense and hypodense areas and uniform hyperdense lesions. Chen (9) reported MR demonstration of aggressive fibromatosis of the tongue. It showed

the mass to be mildly hypointense on T1-weighted image and minimally hyperintense on T2-weighted image. Aisen (10) reported two cases of this condition by MRI study, the lesions were low signal intensity on both images sequence.

In conclusion, the imaging by ultrasonography, angiography, CT scan and MRI study of this condition is not specific and biopsy is needed for the definitive diagnosis.

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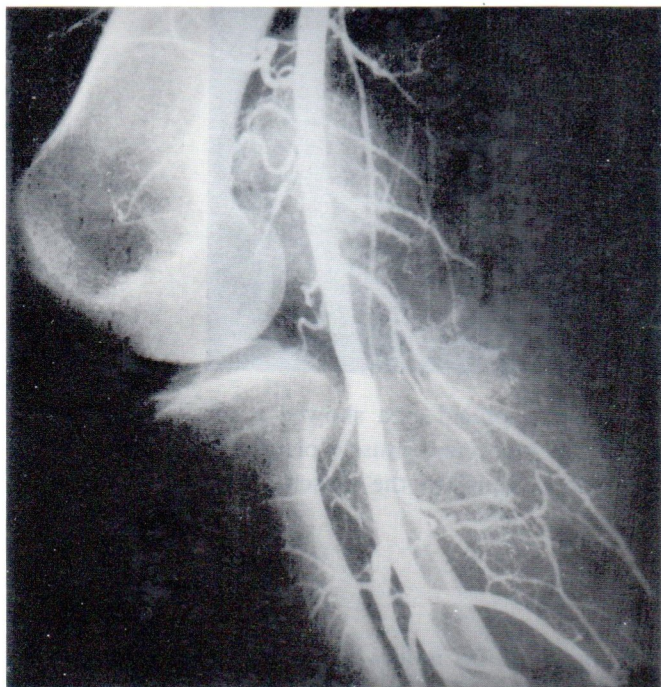


Fig. 2B. Case 2. Right femoral angiography showed two lobulated masses at right popliteal fossa with faint neovascularity and tumor stain.

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Fig. 3A Case 3. MRI study of the mass at posterior aspect of left thigh showed mixed signal lobulated mass at the semimembranosus muscle. The signal included dark grey, light grey, faintly bright on T1WI and dark grey, light grey, faintly bright and very bright on T2WI.

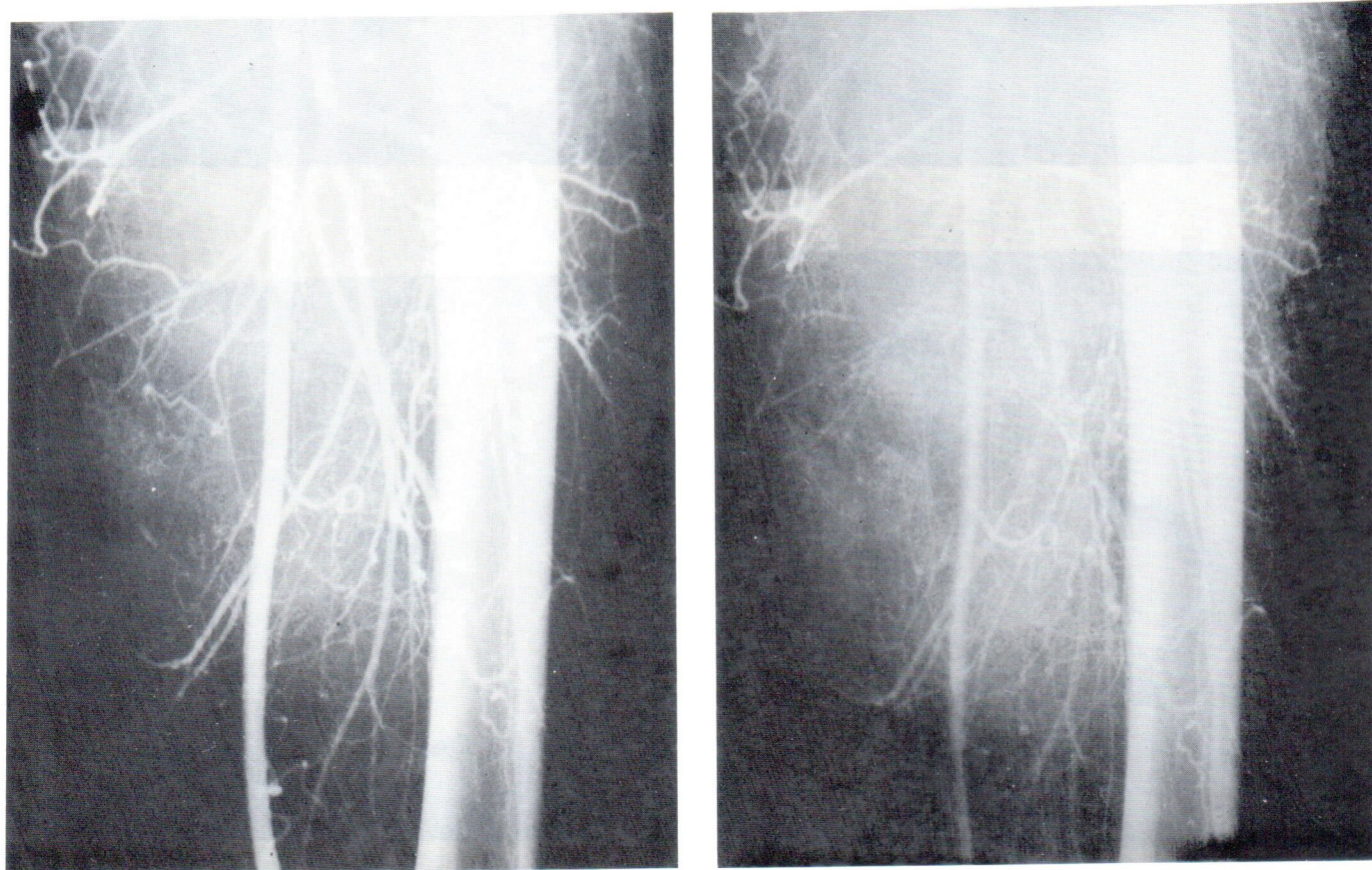


Fig. 3B Case 3. Left femoral angiography showed ill defined border lesion with fainter tumor stain than the tumors in case 2.

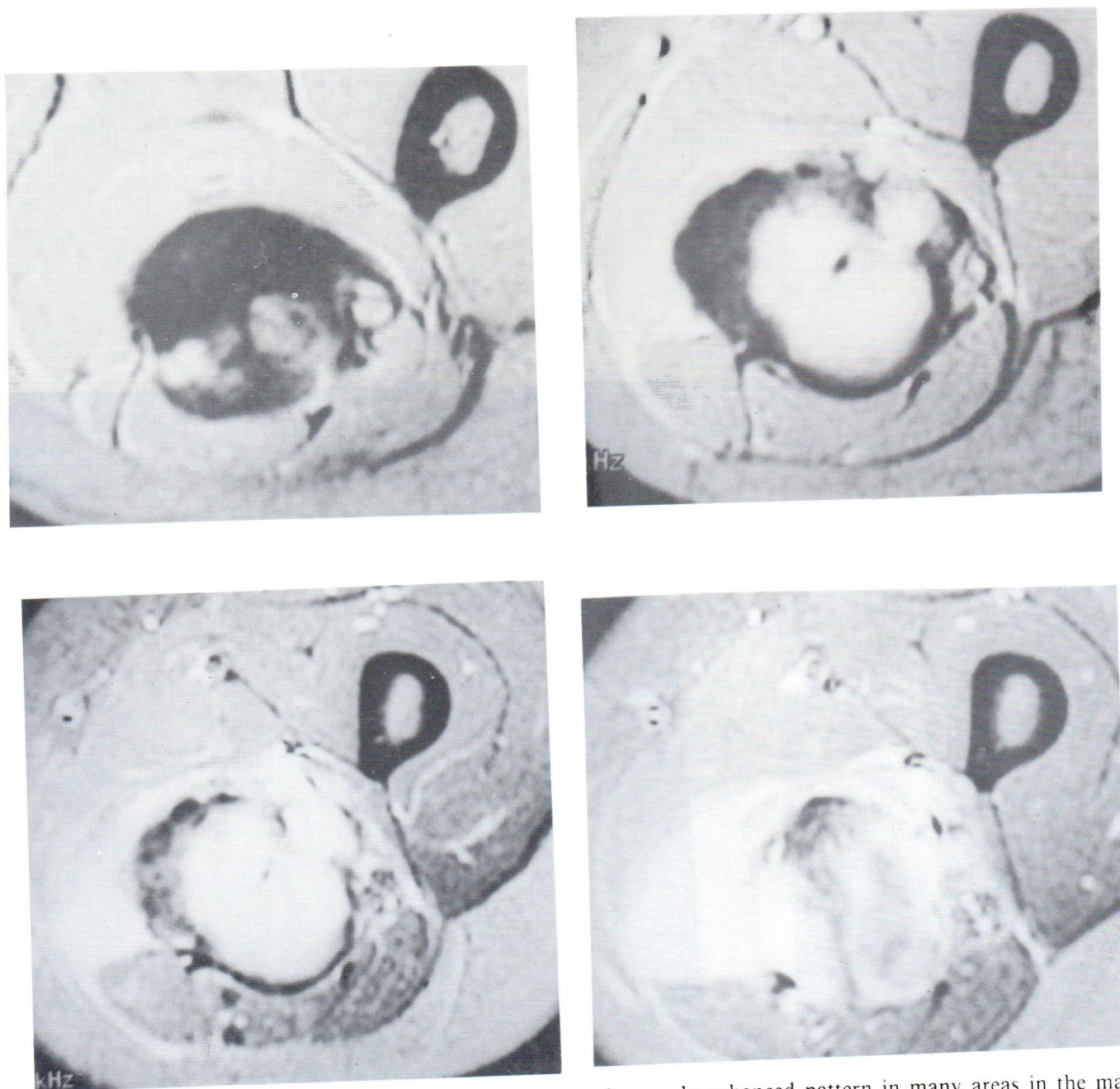


Fig. 4A. Case 3. T1WI axial view post Magnevist showed strongly enhanced pattern in many areas in the mass and the surrounding tissue.

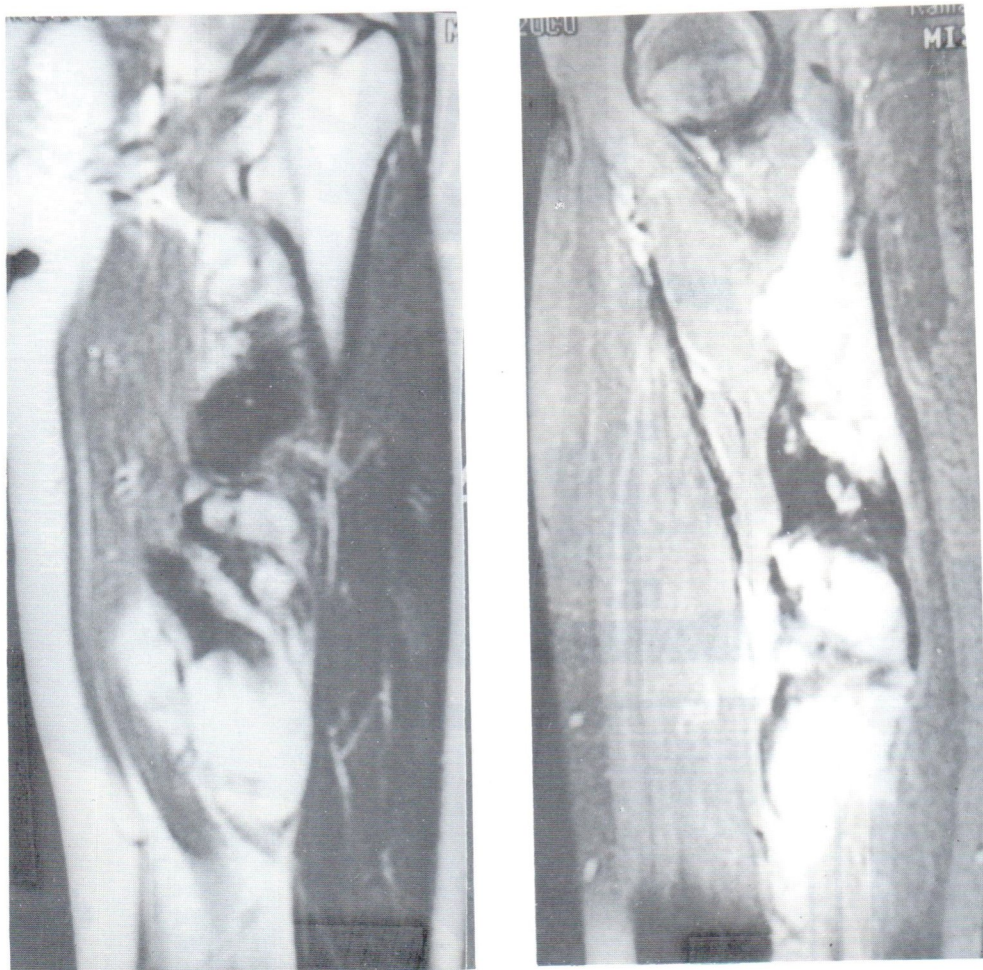


Fig. 4B, Case 3. Post contrast enhancement MRI of the lesion in coronal and sagittal view.