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Case Report

Giant fibrosarcoma of anterior abdominal wall: a rare case report and review literature

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ABSTRACT

Adult type fibrosarcoma is very rare malignant soft tissue tumor. It usually arises in soft tissues of extremities, trunk, head and neck. Fibrosarcoma is essentially a diagnosis of exclusion from other spindle cell mesenchymal malignant neoplasm; by definition negative for epithelial, myogenous and neural markers on immunohistochemistry. To the best of our knowledge, very few cases have been reported in the literature. We here report a case of giant fibrosarcoma arising from anterior abdominal wall in a recurrent dermatofibrosarcoma proturbens and managed with surgical excision and reconstruction with meshplasty.

Keywords: Giant fibrosarcoma, Adult fibrosarcoma, Abdominal wall

INTRODUCTION

Adult Fibrosarcoma (FS) is very rare and it constitutes approximately 1% of adult sarcomas. It is a malignant or intermediate (rarely metastasizing) tumor, composed of fibroblasts with variable collagen production. Fibrosarcomas usually involve the deep tissues of the extremities, trunk, head and neck.[1,2] Adult FS usually appears in the fourth to sixth decades of life with a male predominance.[3,4] Adult fibrosarcoma may arise in dermatofibrosarcoma proturbens (DFSP), Solitary Fibrous Tumor (SFT) and in well differentiated liposarcoma (LPS), either in the primary or in a recurrence, as a reflection of tumour progression.

CASE REPORT

A 55 years male patient presented with a large ulceroproliferative swelling over anterior abdominal wall for 6 months. Physical examination revealed a 15 x 15 cm large lobulated ulceroproliferative growth at umbilical region of anterior abdominal wall (Figure 1). Previously he was operated elsewhere twice for small nodular lesions over same site 5 years & 3 years back respectively and biopsy was suggestive of DFSP.

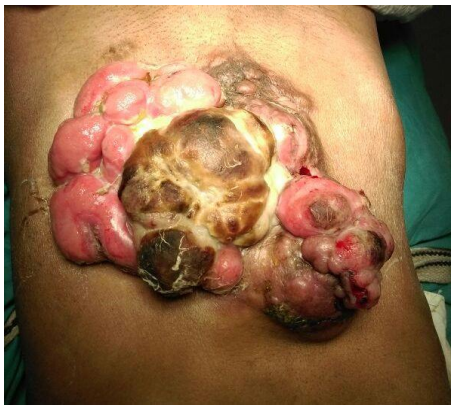

Figure 1: Clinical photograph showing 20 x 20 cm, well-circumscribed mass lesion originating from the anterior abdomen wall.

Contrast Enhanced Computed Tomography (CECT) of abdomen revealed a heterogeneously enhancing soft tissue lesion measuring 16 x 14 x 8.9 cm in size with necrotic areas involving infiltrating skin and muscular plane without intra peritoneal extension (Figure 2a & 2b). CT thorax was negative for distant metastasis. True cut biopsy from the mass was suggestive of high grade sarcoma. In view of no major intra-abdominal extension and preoperative diagnosis of sarcoma, the patient was planned for surgery.

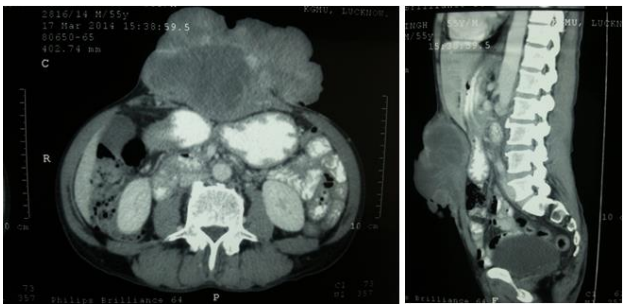

Figure 2a & 2b: Computed tomography (CT scan) showed a heterogeneously enhancing soft tissue lesion measuring 16 x 14 x 8.9 cm in size with necrotic areas involving infiltrating skin and muscular plane without intraperitoneal extension.

Patient underwent wide local excision of soft tissue mass with full thickness of abdominal wall (Figure 3). Reconstruction with composite vicryl and prolene mesh was done. Skin was closed primarily in midline over mesh after lateral flank release incisions (Figure 4a & 4b). Split thickness skin grafting over flank done. On gross examination, cut surface showed solid white lobulated growth measuring 19.5 x 13 x 8.5 cm in size (Figure 5a & 5b).

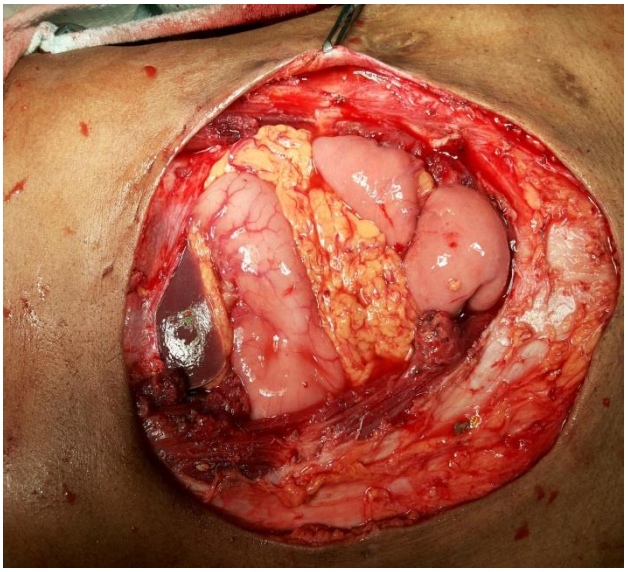

Figure 3: Intraoperative photograph showing a large anterior abdominal wall defect after excision of tumor.

Figure 4a & 4b: Reconstruction of the defect with mesh and primary closure of skin in midline over mesh after mobilization and release incisions [covered with split thickness skin graft (STSG)] in the bilateral flank.

Figure 5a & 5b: Surgical resection specimen showing large ulcereroproliferative and lobulated growth measuring 19x 13.5x 9 cm.

Microscopically, tumour showed spindle cells arranged in sheets and intersecting fascicles. The cells were plump to oval with pale with poorly delimited cytoplasm and moderately pleomorphic elongated tapering nuclei along with frequent mitoses, focal areas of necrosis and myxoid changes (Figure 6). No definite evidence of vascular permeation, perineural invasion or lymphatic permeation was seen. Resection margin of the swelling were clear.

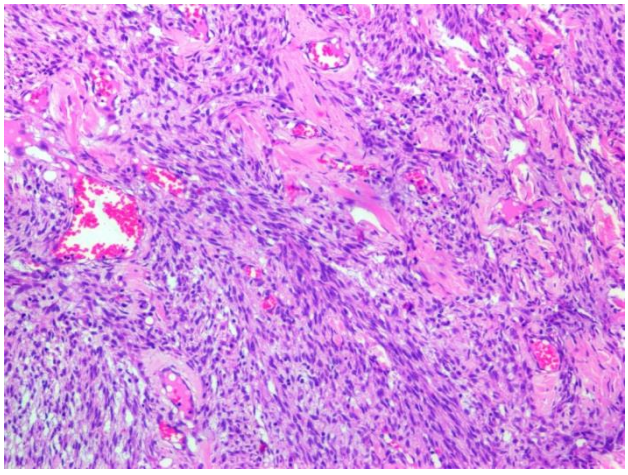

Figure 6: Photo microscopy showing spindle cell tumor with plump to oval shaped cells arranged in sheets and intersecting fascicles.

Immunohistochemical studies revealed that spindle cells were strongly positive for vimentin (Figure 7a) and negative for SMA, CD-34, S-100, desmin and CD-117 (Figure 7b to 7f). Patient received adjuvant chemotherapy

(6 cycles of ifosfamide and doxorubicin) and adjuvant radiotherapy. No recurrent lesion was found using abdominal ultrasound (USG) examination and CECT scan at 15 months after completion of treatment.

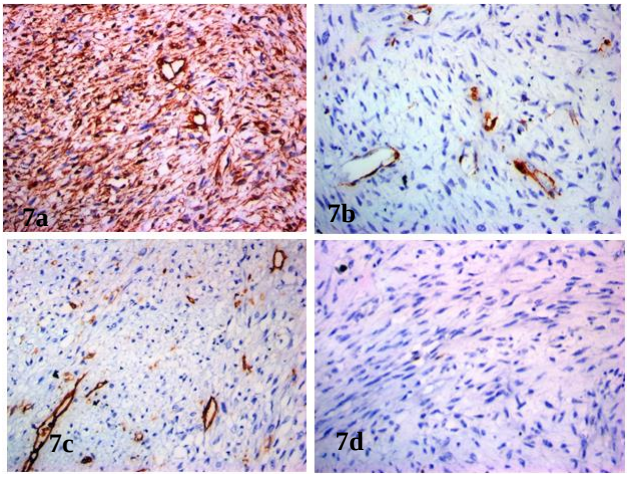

Figure 7 (a-d): Immunohistochemical analysis revealed epithelial elements showed strong positivity for vimentin (7a), negativity for SMA (7b), CD-34 (7c), S-100 (7d).

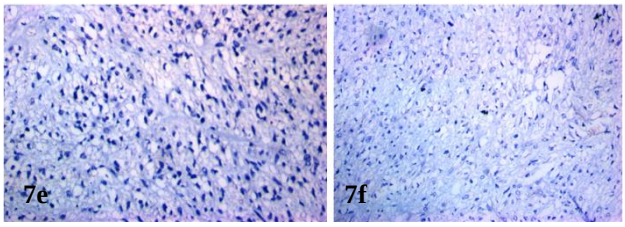

Figure 7e & 7f: Immunohistochemical analysis revealed epithelial elements showed strong negativity for desmin (7e) and CD-117 (7f).

DISCUSSION

Malignant soft tissue tumours of the abdominal wall consist of desmoid and non-desmoid soft tissue sarcomas. They tend to invade adjacent musculo-aponeurotic and bony structures, Transperitoneal organ invasion can also occur with devastating sequelae, especially after an incomplete excision.[5]

The World Health Organization (2002) defined fibrosarcoma as a malignant tumor, composed of fibroblasts with variable collagen and, in classical cases, it have a herringbone bone pattern on light microscopy.[6] Conventional fibrosarcoma falls into two main groups, the adult and infantile types, both very uncommon. Once considered the common adult sarcoma, the incidence of adult fibrosarcoma has declined dramatically over the past several decades. This is due to (i) evolution in the classification of soft tissue tumours (ii) recognition of clinically, morphologically and genetically distinctive subtypes of fibrosarcoma and (iii) increased understanding of the many other mesenchymal and non-mesenchymal tumours that may mimic fibrosarcoma.[7]

Bahrami et al. reevaluated 163 cases originally diagnosed as adult fibrosarcoma from institutional achieves between 1960 and 2008. Revised diagnoses were made on clinical, morphologic, IHC and molecular findings. Among them 84% cases were classified as other than fibrosarcoma. They concluded that true FS is exceedingly rare, accounting for <1%.[4]

Adult FS is essentially a diagnosis of exclusion from other spindle cell mesenchymal tumors; by definition, it is negative at the immunohistochemistry stains for epithelial, myogenous and neural markers, as well as for CD34, CD99, bcl-2 and nuclear beta-catenin.[2,8,9] In the present case, spindle cells were positive for vimentin while negative for all other markers including CD-117, SMA, CD-34, S-100 and desmin.

Fibrosarcomas metastasize to lungs and bone, especially the axial skeleton, and rarely to lymph nodes. Metastasis occurs in 9-63% of cases and 5 year survival rate 39-54%[2]. Surgery remains the principal therapeutic modality in soft tissue sarcoma. Post-operative external beam radiotherapy is helpful to reduce the likelihood of local recurrence in high grade sarcoma.

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Abbreviations

FS - fibrosarcoma, DFSP - dermatofibrosarcoma proturbens, CECT - contrast enhanced computed tomography, CD - cluster of differentiation, SMA-smooth muscle antigen, IHC - immunohistochemistry.

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