

Retroperitoneal Evans Tumour

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Abstract: Here we describe a rare case of retroperitoneal low grade fibromyxoid sarcoma presenting with unusual enlargement of abdomen and subsequently posted for excision but intraoperatively found to have inoperably huge mass completely filling abdominal cavity and could do only debulking surgery.

Keywords: retroperitoneal low grade fibro myxoid sarcoma.

INTRODUCTION

Low grade fibromyxoid sarcoma is a very rare, cytologically bland malignant neoplasm with alternating fibrous and myxoid stroma of low-grade/low malignant potential which was described first by Evans. Diagnosis of LGFMS remains problematic because of its bland looking histologic features that can be potentially confused with other benign or low grade soft tissue tumors. Here we report case retroperitoneal low grade fibromyxoid sarcoma which presented with unusually enlarged abdominal mass.

CASE REPORT

A 45 yr old male presented with progressive abdominal distension, loss of weight and appetite of 3 years duration. Physical examination revealed emaciation and uniformly distended massive abdomen,

with dilated veins over abdomen and lower chest with direction of flow from below upwards (Figure 1). No fluid thrill or shifting dullness was present. Computed Tomography (CT) of abdomen showed a large intraabdominal, intraperitoneal heterogeneously enhancing mass lesion almost filling the whole abdominal cavity, with high and low attenuating areas with septations and calcification displacing bowel posteriorly (Figure 2). CT chest was normal. A CT guided biopsy was suggestive of low grade fibromyxosarcoma. Patient was posted for surgical excision of the abdominal mass and only surgical debulking was possible due to inoperability of mass intraoperatively. The patient was succumbed to death few days after surgery due to severe sepsis and surgical site oozing.



Fig-1: Clinical image of patient with massive distension of abdomen

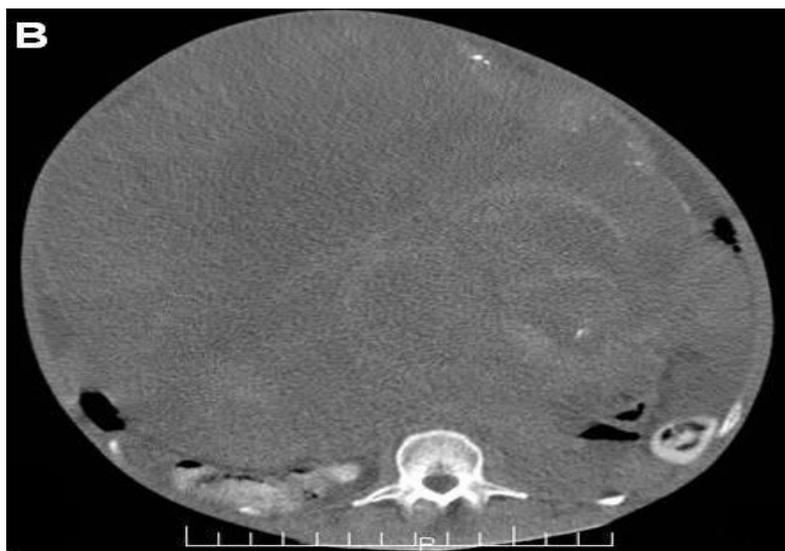


Fig-2: Computed Tomography (CT) of abdomen showing large intraabdominal mass lesion almost filling the whole abdominal cavity, with septations and calcification displacing bowel posteriorly

DISCUSSION

Low grade fibromyxoid sarcoma is a rare, low-grade malignant soft tissue neoplasm with a potential for local recurrences as well as distant metastases [1]. Low-grade fibromyxoid sarcoma (LGFMS) was first described by Evans in 1987 [2]. They usually presents as a painless mass, typically in the proximal extremities .LGFMS is also known as Evans tumour [3]. These tumours are classically situated in the trunk and lower extremities. The common sites are shoulder, thigh and inguinal. Abdominal location is extremely uncommon in which the few cases published in the literature are characterized by slow tumoral progression and long recurrence-free intervals [4].

Differential diagnosis of LGFMS includes lesions showing spindle cell proliferations with myxoid pattern with or without fibrous component, desmoid tumour, desmoplastic fibrosarcoma, and low-grade differentiated liposarcoma. The entities with predominantly myxoid pattern without significant fibrous component include myxomas, low-grade myxo fibro sarcoma, angio myxomas, myxoid liposarcoma, and a myxoid neurofibroma. Tumours with mixed myxoid and fibrous morphologies include neurofibroma, fibromatosis, perineurioma, malignant peripheral sheath tumour, and fibrous histiocytoma. Diagnosis of LGFMS is still difficult because of its characteristic bland-looking histologic features that can be confused with other benign or low-grade fibromyxoid lesions. However, recent cytogenetic and molecular analyses have provided significant

improvements in the diagnosis of LGFMS. Such analyses have demonstrated that most cases of LGFMS have a characteristic t (7, 16) (q33; p11) translocation, resulting in the FUS-CREB3L2 fusion gene. When radically excised, the prognosis is usually good. Radical excision of LGFMS is the cornerstone of treatment. Best treatment for locally recurrent disease is re-excision .No role for chemotherapy has been described.

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