Case Report

Giant omental inflammatory myofibroblastic tumour causing intestinal obstruction: a rare care report and review of literature

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ABSTRACT

It is a rare mesenchymal tumour of intermediate biologic potential (according to World Health Organization), with unknown aetiology. It is benign tumour with malignant potential. It frequently recurs and rarely metastasizes. Abdominopelvic inflammatory myofibroblastic tumours have the recurrence rate of 85% so meticulous follow up is necessary. Complete surgical excision is the main stay treatment.

Keywords: Inflammatory myofibroblastic tumour, Giant, Intestinal obstruction, Omentum

INTRODUCTION

Inflammatory myofibroblastic tumour (IMT) is one of the rare mesenchymal tumours with uncertain biological behaviour. Synonyms of IMT are inflammatory pseudo tumour, inflammatory fibro sarcoma, plasma cell granuloma, pseudo sarcoma, and atypical fibro myxoid tumour. It affects mainly lung but extra pulmonary site has been reported. It is mostly in children and young adult, mostly in second decade of life. Meis et al in 28 cases reported the tumour size ranged from 2.4 to 20 cm (median, 9.1 cm; mean 9.6 cm).1 Coffin et al reported the size from 1cm to 17 cm (mean, 6.4; median,6) in 84 cases, out of these 7 were omental IMT. Omental IMT is rarest tumour among all and to the best of our knowledge about 20 cases have been reported till now in literature.1-5 Singhal et al, Kye et al and Aptel et al reported the similar case.3-5 Here, author reported a rare case of omental IMT of size 40x22x5 cm in 14-year female which present in emergency with acute abdomen and it the largest IMT ever reported in literature till date.

CASE REPORT

A 14-year female patient presenting in our surgical emergency with chief complaints of pain in whole abdomen for 2 months, insidious onset, continuous, diffuse, non-radiating, non-referred, mild to moderate intensity, no aggravating and relieving factors. She also has history of abdominal distension for 1 week and non-passage of flatus and stool for 2 days. No history of fever, vomiting, jaundice, weight loss or loss of appetite. Bladder habit was normal. No history of alcoholic intake. Vital signs were stable. On General physical examination pallor was present. The abdomen was distended with diffuse tenderness present in whole abdomen. A solid palpable lump of size 30x20 cm, smooth, side to side mobility present, ill-defined upper and lateral margin, lower margin not palpable, involving epigastric, right and left hypochondrium right umbilicus, hypogastric, right and left lumber and right and left iliac fossa. No hepatosplenomegaly. On percussion there is dullness present over the lump. Bowel sound absent.
Routine blood tests including a complete blood cell count (CBC) and chemistry studies had normal results with haemoglobin of 8.6 mg/dl and WBC 9600/mm³. ABG was normal. An abdominal ultrasonography shows ill-defined hypo echoic mass, displacing the bowel loop poster lateral occupying whole abdomen. CECT abdomen and pelvis suggested of smudging with increased density stranding is seen in mesentery and along the omentum. Omental and peritoneal thickening showing enhancement on CECT is noted (pre contrast 25-30 HU value and post contrast 38-45 HU value). Moderate ascites is present.

Patients underwent exploratory laparotomy and tumour excision with omentectomy done.

Intraoperative finding suggestive of lobulated, non-capsulated yellowish white firm and gelatinous mass of size 40×20 cm, present in peritoneal cavity with focal Omental involvement. Bowel loops are collapsed. No deposits present over liver, spleen and bowel loops. Bilateral ovaries and uterus are normal. Peritoneal fluid sends for C/S, malignant cytology and specimen for histopathological examination.

Aspirated fluid cytology examination revealed no malignant cells.
Histopathological features suggested of mesenchymal tumour. Most likely Inflammatory myofibroblastic tumor. Post-operative course was uneventful and patient discharge on POD- 7 and refer to medical oncologist for further management. There Immunohistochemically, the tumour cells were positive for vimentin and smooth muscle actin and negative for desmin, S100 protein, CD117, anaplastic lymphoma kinase.

**DISCUSSION**

Inflammatory myofibroblastic tumour are the benign tumour with some rare malignant potential, affected paediatric population with mean age of 10 years and with female predominance, rare after the age of 30. Omental IMT is the rarest tumour. Singhal et al reported the similar case of multifocal intraabdominal IMFT involving omentum and mesentery mimicking peritoneal carcinomatosis in 15-year female presented with intermittent fever, abdominal pain, and weight loss. Aptel et al also reported the ultrasound and CECT finding of IMT of size 6x6x7 cm arise from greater omentum and confirmed by histopathology. The etiopathogenesis of inflammatory myofibroblastic tumour is not well known. Some response to tissue injury, trauma, previous surgery or infection may be the cause.

IMT is also known by the inflammatory pseudo tumour, inflammatory fibro sarcoma, plasma cell granuloma, pseudo sarcoma, and atypical fibro myxoid tumor. IMT was first reported in lung in 1939 by Brunn. Inflammatory pseudo tumour of the lung, is the most common organ effected in children younger than 16 years of age and most common extra pulmonary organs are the liver, spleen, head and neck, heart and retroperitoneum. Some literature also reported Chromosomal abnormality, ALK-1 gene, and autoimmune disease, translocation of long arm of chromosome no 2 and short arm of chromosome no 9 may be the other cause. According to World Health Organization it is a neoplasm of intermediate biologic potential because of local recurrence and distant metastases. Differential diagnosis of Abdominopelvic IMT are lymphoma, sarcoma, desmoids tumour, carcinoid tumour, and gastrointestinal stromal tumor. Most common presenting symptom is the pain in abdomen but may present with vague abdominal lump, anorexia, weight loss and obstructive features. Recurrence rate is about 85% in abdominopelvic IMT; most of the patients have the recurrence in first year of surgery. Some patients show the recurrence up to 9 year of first surgery. Treatment of recurrence is re-excision of tumour or metastasectomy. Radical surgery should not be attempted as it causes high morbidity and mortality, only simple excision is sufficient for the IMT. So, complete surgical excision with R0 resection is the treatment of choice with good prognosis, although some case reports of spontaneous regression have been found in literature. Some patients also show response to steroids, radiotherapy, chemotherapy and even NSAID's like ibuprofen. Anaplastic lymphoma kinase reactivity
show favourable prognostic factors and chemotherapy and radiotherapy is preserved for the cases that recurs or metastasize. Due to local invasiveness and recurrence tumour are indistinguishable from malignancy so, proper preoperative workup to prevent radical treatment and regular follow up with radiological investigation is necessary in these patients.

CONCLUSION

We conclude that abdominopelvic IMT is rare mesenchymal tumour, affecting mainly paediatrics group female, with unknown aetiology. Surgery is the main stay treatment but reoccurrence is very common. Proper knowledge regarding the tumour prevents the radical treatment and reduces morbidity and mortality.

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