Abdominal Inflammatory Myofibroblastic Tumor with early recurrence: A case report analysis

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ABSTRACT

Inflammatory Myofibroblastic tumour (IMT) is a rare benign disease of unknown etiology and pathogenesis with a potential of local infiltration & multicentricity. This rare tumour occurs primarily in the lungs, but has occurred in other extra-pulmonary sites. We report the case of a 9 year old girl presenting with abdominal mass, weight loss, fever, easy fatigability & malaise for one month. Imaging showed large lobulated heterogeneous mass of 7x11 cm with necrotic & calcified foci & fatty liver. She was operated with intraoperative findings of large multilobular cyst arising from omentum and transverse mesocolon near the splenic flexure. Biopsy and immunohistochemistry (IHC) confirmed the diagnosis of IMT. Unfortunately, within a period of three months she had recurrence hence was put on chemotherapy but there was no regression of disease. It was decided to re operate her. She is doing well except one episode of febrile neutropenia and is on our follow up.

KEYWORDS: Inflammatory Myofibroblastic tumour, Early recurrence

INTRODUCTION

Inflammatory myofibroblastic tumor (IMT) is a relatively new histopathologic term describing an entity previously known as an inflammatory pseudotumor. They can be found virtually at any anatomic site, with a predilection for the lung, the genito-urinary tract and the mesentery [1]. It was first observed in the lung and described by Brunn in 1939 and was so named by Umiker et al. in 1954 because of its clinical and radiological behavior that mimics a malignant process [2]. The disease was first described in 1984 but the disease etiology remains unknown [3]. The whole mark of these tumors is the spindle cell proliferation, which are in fact myofibroblasts, associated with a variable inflammatory component and hence the name [4]. Previously believed to have an inflammatory or reactive pathogenesis, are now viewed as a neoplasm due to a propensity of local recurrence and metastasis [5].

Herein, we present a case of IMT of the mesentery who presented clinical and radiological findings of malignancy and was diagnosed after histopathological and immunohistochemical evaluation of the resected tumor.
CASE REPORT

Our case was a 9 year old girl presenting with abdominal mass, weight loss, fever, easy fatigability & malaise for one month. On examination patient had mild pallor, weight of 20 Kg and a large 11x7 cm firm immobile swelling was palpable in left upper abdomen. Cardiovascular system, central nervous system, respiratory system & other general physical examination was normal. Investigations revealed Hb of 9gm/dl, thrombocytosis (5 lack/mm$^3$), C - reactive protein 230 mg/dl and elevated ESR (30mm /hr). Imaging showed large lobulated heterogeneous mass of 7x11 cm with necrotic and calcified foci & fatty liver. With this background gastrointestinal stromal tumour (GIST), castle man’s disease, lymphoma, renal or other abdominal malignancy was suspected.

She was operated with intraoperative findings of large multilobular cyst arising from omentum with multiple adhesions to transverse colon and another solid mass of 7x6 cm arising from transverse mesocolon near the splenic flexure. Complete excision was done. HPE examination of the specimens revealed spindle cell tumour with marked inflammatory background of lymphocytes, plasma cells, neutrophils, with minimal mitosis & absent atypia. Preliminary diagnosis of inflammatory myofibroblastic tumor was made. IHC confirmed that with strong positivity for desmin, SMA & ALK-1 and negativity for CD-34 & CD-117, so diagnosis of IMT was made.

Unfortunately, within a period of three months only patient again developed earlier symptoms with pain abdomen. Repeat imaging showed two heterogeneous masses; large one in left upper abdomen & small one in pelvis. She was put on chemotherapy based on cisplatin & doxorubicin. She received three cycles but with no regression of disease. It was decided to re operate her. Intraoperative findings were huge friable mass arising from mesentery of jejunum about 10 cm from DJ junction. Tumor was adhered to the adjacent jejunal segment and there was another mass 4x5cm in the pelvis arising from sigmoid mesocolon. Specimens from both masses were again put to HPE which were in confirmatory of the earlier diagnosis. So diagnosis of IMT with a recurrence was made. Patient is on our follow up. She is doing well except one episode of febrile neutropenia. Repeat USG is normal except for fatty liver.

DISCUSSION

Inflammatory Myofibroblastic tumor is a benign condition [6]. IMT has been described by various names including inflammatory pseudotumor (IPT), plasma cell granuloma, benign myofibroblastoma, inflammatory fibrosarcoma, histiocytoma, xanthomatous granuloma, and spindle cell pseudotumor, describing its heterogeneous nature [7]. It can be multicentric as in our case and has a potential to locally infiltrate [8]. It is commonly found in lung. Other less common sites being mesentery, omentum, liver, retroperitonium etc. but can occur anywhere in the body [1,9].

They mainly involve the lungs of children, without sex predilection, whereas, extra-pulmonary locations are manifested mostly in the viscera with a slight predominance in females [10].It sometimes mimic commoner conditions like sclerosing inflammatory lesions, nodular fasciitis, lymphomas, fibromatosis, GIST, inflammatory fibroid polyps and Castleman’s disease. Though many etiologies like HH8 & EBV viruses, IL-6 over expression etc have been suggested but none has been proved beyond doubt [11,12].

Lawrence et al. identified two distinct balanced chromosomal translocations involving the ALK kinase gene. One at chromosome 2p23 and other involving either the tropomyosin 3 gene (TPM3) at chromosomal locus lq21 or the tropomyosin 4 gene (TPM4) at chromosomal locus 19p13.1, resulting in t(1;2) (q21;p23) and t(2;19) (p23;p13.1) translocations, respectively [13]. ALK positivity has been detected in 40% of IMTs [1]. Symptoms depend upon the site of tumour location. Some common symptoms being swelling, fever, easy fatigability, weight loss, pain, decreased appetite (as was in our patient), dyspnea, chest pain, haemoptysis etc. [1].
Laboratory investigations usually suggest an inflammatory process: leukocytosis, neutrophilia, elevated ESR and C-reactive protein (CRP), anemia, thrombocytosis and polyclonal hypergammaglobulinemia [14]. Imaging usually show heterogeneous masses with necrosis as depicted in Fig 1 & 4.

Figure: 1 Preoperative CT image showing a large heterogenous enhancing mass with central necrosis & foci of calcifications measuring 11x7 cm (arrows)

Figure: 2 Low power view of HPE showing spindle cells with inflammatory background of plasma cells, lymphocytes & neutrophils

Figure: 3 High power view of HPE showing spindle cells with inflammatory background of plasma cells, lymphocytes & neutrophils

Histologically, IMT corresponds to a fascicular proliferation of spindle cells (fibroblasts, myofibroblasts), intermingled with a chronic inflammatory infiltrate (plasmocytes, lymphocytes, histiocytes and some eosinophilia) and a fibrous connective tissue. There is neither nuclear atypia nor mitoses [15] as shown in Fig 2 & 3.

Immunohistochemically, the myofibroblastic spindle cells can be positive for vimentin (99%), smooth muscle actin (92%), muscle-specific actin (92%), desmin (69%), cytokeratin (36%), CD68 (KP-1) (24%), and CD30 (Ki-1) (6%).[1] Surgical excision is the treatment of choice. Radical excision is curative in more than 90% of extrapulmonary IMTs [16].
Incomplete excision, extra pulmonary location, size greater than 3cm, ill defined margins, ganglion like cells cell, atypia, p53+, round cell morphology & nuclear membrane staining of ALK favor recurrence [17,18]. Recurrence rates vary according to anatomical site, being < 2% for tumours confined to the lung, increasing up to 25% for extra pulmonary lesions [19]. A favorable response to radiation therapy occurs in 75% of patients. Corticosteroids are generally not useful in adults, although good results have been reported in children in cases of unresectable tumors or hilar and mediastinal invasion. Chemotherapy is useful in cases of multifocal, invasive lesions or in cases of local recurrence [20]. With complete surgical excision 3 & 5 year survival rate is 85% & 74% respectively. Recurrence is usually seen after one year but in our patient it was only after three months.

CONCLUSION AND RECOMMENDATIONS

Though IMT is a rare tumor we should always keep it as a differential to any lung or abdominal lesion particularly in children. Even with multimodal pre-surgical investigation, it can be extremely difficult to differentiate inflammatory myofibroblastic tumor from other malignancies. Though imaging may help, HPE & IHC are diagnostic. Spindle cells (myoepithelial cells) with inflammatory background are characteristic. Most cases require surgical exploration and complete resection to obtain an accurate diagnosis. Surgical excision is the treatment of choice.

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