ORIGINAL RESEARCH PAPER

Paediatric Surgery

RARE CASE OF OMENTAL INFLAMMATORY MYOFIBROBLASTIC TUMOUR.

KEY WORDS: Inflammatory Myofibroblastic tumour(IMFT), Omentum.

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ABSTRACT

Inflammatory myofibroblastic tumour (IMFT) is a relatively uncommon neoplasm with unpredictable malignant potential known to occur anywhere in the body. IMFT involving the omentum is a very rare entity with less than 15 cases reported so far [2,3]. We are presenting a case of 5-year-old presented with complains of fever and pain in abdomen since 2 months. Intraoperative findings were suggestive of a well defined omental mass with no evidence of metastasis or involvement of other structures. Diagnosis of same was confirmed on histhopathology.

INTRODUCTION

Inflammatory myofibroblastic tumour (IMFT) more commonly known as inflammatory pseudotumour is a relatively rare tumour with variable malignant potential. Typically seen in children and young adults it can occur anywhere in the body but is commonly described in the lungs, mediastinum and orbits. Intraabdominal IMFT commonly involves liver, stomach, bowel and spleen. [1]

IMFT lesions microscopically consists of myofibroblastic spindle cells. The tumours contains pro-inflammatory white blood cels and express highly abnormal oncogenic fusion protein mainly anaplastic lymphoma kinase(ALK).

Case Report

Five year old female child presented to our outpatient department with pain in abdomen and fever since 2months. Pain in abdomen was sudden in onset gradually progressive , dull aching in nature, relieved temproryly on medication with no aggravating or relieving factor. Fever was low grade and intermittent in nature. There were no bowel or bladder complains.

On physical examination there were no palpable lump in the abdomen. There was no tenderness or guarding or rigidity on palpation.

Per rectal examination revealed no abnormality.

Patient was admitted in our ward for further investigation for recurrent pain in abdomen.

Patient underwent routine blood investigations along with amylase and lipase level were sent and ultrasonography was done.

Laboratory investigations revealed anemia hemoglobin of 4.2g/dl which was corrected by two blood transfusion with pre operative hemoglobin of 12.1g/dl. Ldh levels were found to be raised(871units/dl). Amylase and Lipase levels were within normal range.

Ultrasonography of abdomen and pelvis were suggestive of a well-defined hypoechoic solid cystic lesion measuring 3*3.6* 4.4 cm noted in right adnexa with right ovary not separately visualized with rest of the ultrasonography showing no other abnormality.It was showing increased internal vascularity with arterial waveform on spectral doppler, and mild free fluid in pelvis without internal echos. Left ovary within normal limits. Features suggestive of neoplastic etiology of right ovarian origin.

Metstatic workup of this patient was negative.

MRI abdomen and pelvis was suggestive of well defined T1 iso intense, T2 3.3* 3.5*4.2 cm sized well defined hyperintense solid lesion with restricted diffusion.

Anteriorly it was related to lower abdominal wall and loops of small bowel. Posteriorly it was abutting the right psoas muscle & right external iliac artery. Medially abutting small bowel loops. Laterally it was in close relation to caecum. No evidence of involvement of other organs on MRI was seen.

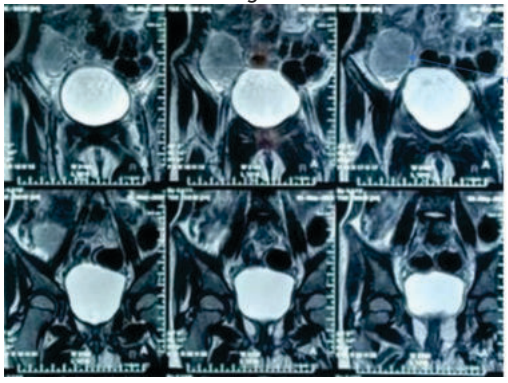

Figure 1- MRI Abdomen Pelvis

MRI Abdomen and Pelvis is showing a T2 hyperintense solid mass with restricted diffusion

Patient underwent evaluation under anesthesia. The mass was palpable on deep palpation under anasthesia. The mass was freely mobile in all directions. Lump was firm in consistency.

The decision to do a diagnostic laparoscopy was taken. 5mm Umbilical camera port and two 5mm working were inserted in left and right lumbar region.

Intraoperative findings were suggestive of mass of size about 3.3*3.5*5 cm, freely mobile in all direction and the mass was arising from the omentum. The mass was free from both ovaries and uterus. No adhesion were seen between the mass and and bowel wall.No other organs were involved. No obvious lymph nodal enlargement seen.

Laparoscopic excision of omental mass was done and the specimen was sent for histopathological examination. Complete bowel walk was done and there was no evidence of other lesions in the abdomen or pelvis.

Histopathological examination was suggestive of low grade spindle cell neoplasm with focal haemangio pericytomatous patter. Immunohistochemistry done with IHC marker ALK1 & SNA was positive whereas CD23 & c KIT were negative.

Figure 2 Intraoperative picture suggestive of relation with various organs As seen in the diagram the mass is seen arising from omentum and is free from all surrounding structures. Minimal free fluid seen in pelvis

DISCUSSION

IMFT is a rare tumor of intermediate biologic potential as regards malignancy as described in the recent WHO classification. It has been variously called inflammatory pseudotumour, inflammatory fibrosarcoma, plasma cell granuloma, pseudosarcoma, and atypical fibromyxoid tumour. [2]. Initially described in the lungs and orbit as a reparative post inflammatory process, it has now been reported in many other extrapulmonary sites. The pathogenesis of this condition has been variously described as an excess inflammatory response to infection, trauma, and surgery. The abdomen is a relatively uncommon site for IMFT; the liver is the organ most often involved. It can occur in both genders and is seen in children and adolescent.

Radiologic findings of omental-mesenteric IMFT are indistinguishable from other malignant lesions. On sonography they are seen as solid iso- to hypoechoic masses with vascularity on Doppler. On CT they are variously described as homo- or heterogeneous hypervascular lesions showing moderate to intense enhancement. Calcification, hemorrhage and necrosis are reported in a few cases [4]

Features suggesting more aggressive behavior include multifocal tumors, retroperitoneal location, infiltration of adjacent structures and incomplete excision. Pathologic features such as tumor cellularity, mitosis and necrosis do not correlate with outcome. Recently IHC marker ALK positivity has been shown to predict lower risk of metastasis [5]

Omental IMFT has been described as a distinct entity from the common inflammatory pseudotumour of the abdomen as pathologically it represents a form of clonal expansion rather than a postinflammatory response [1]

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