Inflammatory Myofibroblastic Tumor of Greater Omentum – A Rare Cause of Intra-abdominal Lump

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Abstract

Inflammatory myofibroblastic tumors (IMTs) are uncommon mesenchymal tumors. These are seen more in children & young adults. These tumors can occur in any part of the body, like in lung, mesentery and omentum. Clinical presentation depends on the location of the tumor with associated low-grade fever, growth failure, malaise & weight loss. IMTs may be multicentric, have a high local recurrence rate. Usually benign, malignant transformation is very rare and may metastasize in rare cases. These lesions show wide variability in their histopathologic features like inflammatory infiltration, predominantly of plasmatocytes & lymphocytes and occasionally neutrophils & eosinophils. Owing to the rarity of these lesions, there are no specific imaging findings that distinguish IMTs from other mesenteric masses. Complete surgical excision is the treatment of choice. Local recurrences are high and re-excision is preferred.

Keywords: Inflammatory myofibroblastic tumors, Inflammatory pseudotumors, Inflammatory fibrosarcomas, Plasma cell granuloma, Mesenchymal tumors

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Introduction:

Inflammatory myofibroblastic tumors (IMTs), also known as inflammatory pseudotumors, inflammatory fibrosarcomas and plasma cell granuloma, are uncommon mesenchymal tumors composed of

myofibroblastic spindle cells admixed with lymphocytes, plasma cells and eosinophils.[1] These are most common in children & young adults.[2] These tumors can occur in any part of the body, most commonly in the lung, mesentery and omentum. Presentation depends upon the location of the tumor. They may also present with associated low-grade fever, growth failure, malaise &weight loss. Patient may be asymptomatic or may present with a painless abdominal lump, or even with only abdominal pain without any palpable lump.[3] IMTs may be multicentric, usually benign and rarely transformed to malignancy. Pre-operative diagnosis is very difficult due to rarity and unusual presentations. Surgery is the treatment of choice.

Case Report:

A 6-year-old girl came to the Surgery outpatient department of Green Life Medical College Hospital with history of recurrent abdominal pain for 2 months. The pain was felt in lower abdomen, of sudden onset, dull aching, moderate in intensity, non-radiating, had no specific aggravating & relieving factor and occurred at recurrent episodes, each episode lasted for 4-5 days. She complained of low-grade intermittent fever, highest peak recorded was 100^0 F. She also had history of recurrent constipation and increased frequency of micturition with sense of incomplete emptying of bladder.

On abdomen examination, an intraabdominal mass was palpable in the right side of abdomen extending both in right lumbar region and right iliac fossa. The mass was non-tender, 5x5 cm, well-defined, with smooth surface, firm in consistency, and was mobile in all directions. There

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was no organomegaly and para- aortic lymph nodes were not palpable.

Ultrasonography of abdomen revealed the presence of a thick walled, well outlined, hazy fluid containing cystic mass measuring about 6cm×5cm in right lower abdomen but did not comment on organ of origin of the mass. CT Scan of Whole Abdomen with Contrast showed a soft tissue mass in the left iliac fossa, with the possible differentials of mesenteric cyst, lymphadenopathy or exophytic gastrointestinal stromal tumor (GIST) arising from bowel loop.

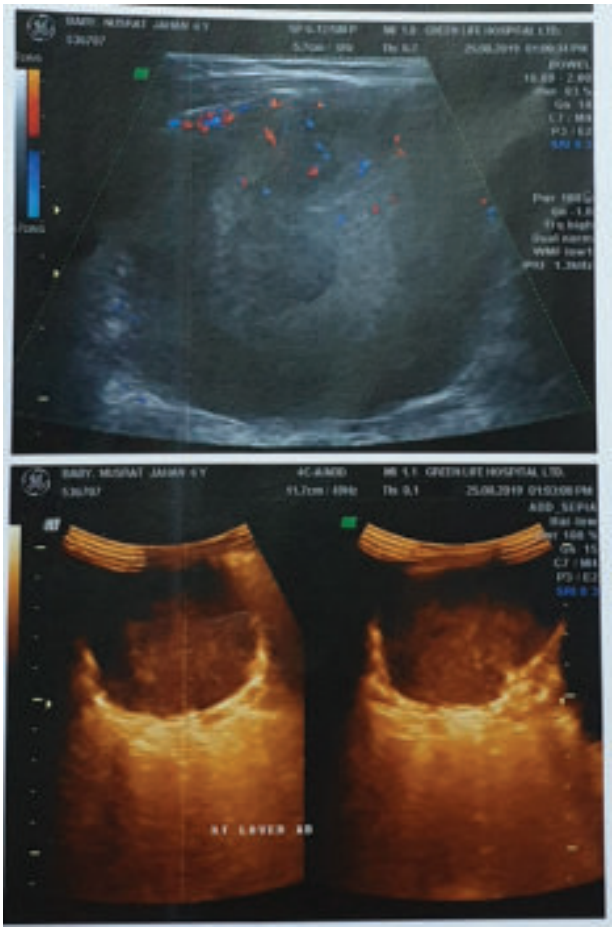

Fig. 1: *USG of whole abdomen showing a thick walled, well outlined, hazy fluid containing cystic mass in right lower abdomen*

Exploratory laparotomy was performed under general anesthesia using a midline incision. A solid lump measuring about 6×5cm was found in pelvic cavity attached to greater omentum by a narrow pedicle. It was not adherent to any other surrounding structures. The lump was excised. All abdominal organs were examined and found to be normal.

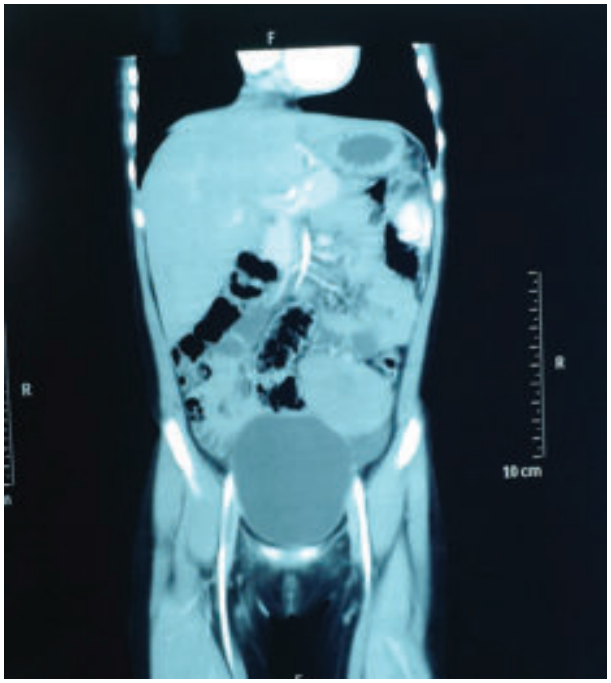

Fig. 2: *Contrast-enhanced CT scan of whole abdomen (coronal section) showing soft tissue mass at the left iliac fossa*

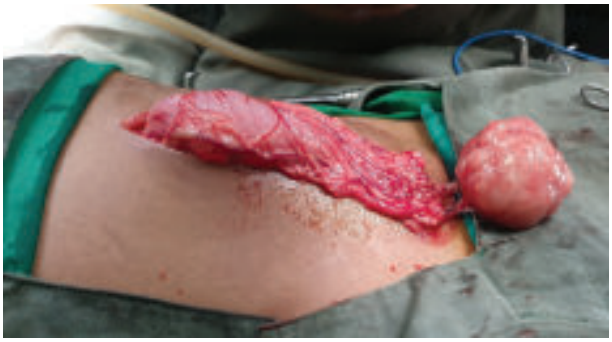

Fig-3: *The lump after laparotomy attached to the greater omentum by pedicle*

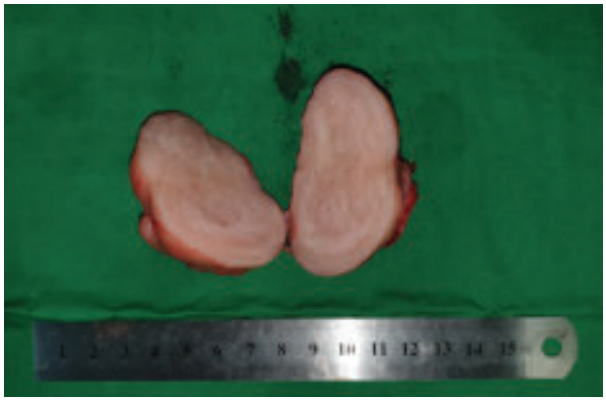

Fig. 4: *Cut surface of the tumor showing fleshy and whorled appearance*

Histopathological examination revealed an inflammatory myofibroblastic tumor made of fibroblasts, many lymphoplasmacytic cells and histiocytes in whorled appearance.

The post-operative period was uneventful. Stitches were removed on 9th post-operative day and she was discharged from hospital. She is now on regular followup.

Discussion:

Inflammatory myofibroblastic tumors(IMTs)are uncommon mesenchymal tumors composed of myofibroblastic spindle cells admixed with lymphocytes, plasma cells and eosinophils.[1] Previously these were thought to be reactive lesions, but nowadays are considered neoplastic.[1] Though these tumors can occur in any organ, but commonly in the lung, mesentery and omentum. IMTs are common in children & young adults with a mean age of approximately 10 years.[2] The etiology of these tumors is unclear. Presentations depend on the location and size of the tumor. Patient may remain completely asymptomatic or present with a painless abdominal lump, or even with only abdominal pain without any palpable lump.[3] Our patient presented with recurrent abdominal pain and a palpable lower abdominal lump which exerted pressure effect on the urinary bladder & sigmoid colon that lead to features of lower urinary tract symptoms& constipation. Macroscopically these firm tumors show white or yellow colored fleshy appearance on cut section with whorled pattern. The masses may be sessile or polypoid. Microscopically these tumors contain a mixture of inflammatory cells and myofibroblasts, and fibroblasts in fascicles or whorls. Immunostaining is positive for smooth muscle actin (86%), muscle-specific actin (82%), desmin(41%), calponin, cytokeratin (26%), and vimentin, factor XIII A, CD68, CD117 may be present in submesothelial areas of the tumor.[4] Cessna *et al* recommended that immunohistochemistry for anaplastic lymphoma kinase (ALK1) and p80 is useful as an indicator of 2p23 abnormality, but it must be interpreted in the context of histologic and other clinicopathologic data if used as an adjunct to differential diagnosis as anaplastic large-cell lymphoma also expresses the same abnormality.[5] Owing to the rarity of these lesions, there are no specific imaging findings that distinguish IMT from other mesenteric masses such as GISTs, mesenteric fibromatosis, sclerosing mesenteritis, fibrohistiocytic sarcomas, Hodgkin and non-Hodgkin lymphoma, and metastatic carcinoma. Ultrasonography determines the solid nature of the lesion, and contrast-enhanced computed tomography (CT) provides the information regarding the shape, extent of involvement and anatomical relationship to adjacent structures. IMTs may be multicentric with a high local recurrence rate. These are usually benign and malignant transformation is very rare; metastasis is even rarer.[6] Treatment of choice is non-aggressive surgical excision of the tumor. Tothova *et al* and Groenveld *et al* recommended radical excision of the tumor as mainstay of therapy due to high local recurrence rate.[7, 8] Re-excision is the treatment of choice for local recurrence and in several studies the majority of such patients remained disease-free with long-term follow up.[2, 9] Chemotherapy and radiotherapy are reserved for progressive disease after complete excision.

Conclusion:

Inflammatory myofibroblastic tumors is a rare tumor. It may present with abdominal pain, mass and pressure effect. Ultrasonography and CT scan are the imaging investigations used to diagnose it. Surgery is the treatment of choice. The patient should be kept follow up to assess recurrence.

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