

Multiple intestinal and extraintestinal inflammatory myofibroblastic tumors-a rare presentation with review of literature

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Abstract: Inflammatory Myofibroblastic Tumor (IMFT) belongs to a rare class of pseudosarcomatous benign inflammatory lesion. It usually affects young adults and children. Lung is the most common site. Mesentery, omentum, liver, spleen, colon are rare sites. We reported a case of 4 year old female child who presented with abdominal distension. On examination multiple masses were seen in small intestine, omentum, omental lymph nodes and few lesions even outside the abdominal cavity in lung. Resected part of small intestine on histological examination showed dense proliferation of spindle cells and inflammatory cells. Case was diagnosed as inflammatory myofibroblastic tumor. The follow-up of the case was uneventful.

Key words: Small bowel, pseudosarcomatous, inflammation

I. Introduction

IMFT of small bowel are rare. Over the last two decades, IMFT has emerged as a distinct entity with characteristic clinical, pathological and molecular features. We described a case of 4 year old girl who have small intestinal resection for inflammatory myofibroblastic tumor with follow up for six months without any clinical problem. IMFT shows a predilection for the visceral soft tissue of children and adolescent and has a tendency for local recurrence, but only a small risk of metastasis. Chromosomal translocation leading to activation of tyrosine kinase can be detected in approximately 50% of IMTs, particularly those that arise in young patients.

II. Case history

A 4 year old girl presented with an abdominal mass of few months duration. She had no other associated symptoms. Obvious clinical swelling of size 8.0x7.5 cms. was palpable on per abdominal examination. The blood investigations showed microcytic hypochromic anemia and increased ESR. An ultrasound of the abdomen revealed multiple masses in abdominal cavity as well as outside the abdominal cavity. In small intestine multiple growths were seen on serosal surface varying in size from 2.3x1.1 to 6.5x6.0 cms. Similar lesions were also seen in jejunum, sigmoid colon, omental lymph nodes and outside the abdominal cavity especially in lung. At lapotomy, the tumor was found to be circumscribed and present in the wall of small and large intestine and also in other areas in abdominal cavity and even outside the abdominal cavity. Affected part of ileum was resected. Patient was followed up for 6 months and was clinically symptomless.

On gross examination, the tumor measured 5.8x5.0x3.0 cms and showed grayish white cut surface. Tumor was well circumscribed (Fig.1). On histopathological examination, tumor was composed of an admixture of proliferating spindle cells and chronic inflammatory cells embedded in variable collagenous or hyalinised stroma. The inflammatory cells were polymorphous, consisted of plasma cells, lymphocytes, eosinophils and mast cells. Mitosis was occasional (Fig 2 & 3). Diagnosis of inflammatory myofibroblastic tumor was made. Immunohistochemistry showed reactivity for vimentin and smooth muscle actin.

III. Discussion

The term inflammatory pseudotumor has been used to describe a wide range of reactive and neoplastic lesions, including inflammatory myofibroblastic tumor (IMFT), pseudosarcomatous myofibroblastic proliferations of the genitourinary tract, infectious and reparative processes and inflammatory pseudotumors of lymph node, spleen and orbit. Over the last two decades, IMFT has emerged as a distinct entity with characteristic clinical, pathological and molecular features.

The term inflammatory myofibroblastic tumor or inflammatory pseudotumor was proposed in 1990. It has been regarded as a benign and reactive disorder for a long time but some cases of IMFT have a malignant course. Inflammatory pseudotumor was first described in the lung, where it was considered a reparative post inflammatory condition rather than a neoplastic process^{1,2}. Similar lesions were reported at extra pulmonary sites including somatic soft tissue, bone, larynx, uterus and central nervous system³. One recent study reported that approximately one third of cases were pulmonary and two third were extra pulmonary⁴. The IMFT affects both sexes, at any ages, with a slight predominance in children and young adults. In our patient, the tumor was discovered at a very early age.

Patient generally present with a mass or non specific symptoms, including vague abdominal pain or gastrointestinal complaints for intraabdominal lesions, and cough, chest pain, or, less often, hemoptysis for pulmonary tumors. A laboratory evaluation may reveal microcytic anemia, a raised erythrocyte sedimentation rate, thrombocytosis, and/or polyclonal hypergammaglobulinemia⁵. An abdominal mass and microcytic hypochromic anemia and raised ESR on hematological investigations were also seen in our case. In some cases, the mass may be found only after an extensive workup for fever of unknown origin or growth failure⁶. The same clinical presentation may be seen in the plasma cell variant of Castelman disease, in which overproduction of interleukin 6 is thought to be the underlying cause of the systemic complaints⁷. Grossly, inflammatory myofibroblastic tumor was well circumscribed but not encapsulated, firm and homogenous with sometime areas of necrosis⁸. In our case, tumor was well circumscribed and showed grayish white cut surface.

Histologically, IMT are characterized by variably cellular spindle cell proliferation in a myxoid to collagenous stroma with a prominent inflammatory infiltrate composed primarily of plasma cells and lymphocytes, with occasional admixed eosinophils and neutrophils. Coffin et al described three basic histological patterns, which are often seen in combination within the same tumor: a myxoid/vascular pattern, a compact spindle cell pattern, and a hypo cellular fibrous pattern. The myxoid/vascular pattern has a fasciitis like appearance, with loosely arranged plump spindle cells in an edematous or myxoid stroma and a prominent vasculature. The inflammatory infiltrate in these areas often contains more neutrophils and eosinophils and fewer plasma cells than in other two patterns. The compact spindle cell pattern is characterized by a cellular proliferation of spindle cells with fascicular or storiform architecture in a collagenous stroma. These foci typically show numerous plasma cells and lymphocytes intimately mixed with the spindle cells, but discrete lymphoid follicles and aggregate of plasma cells are also common. The fibromatosis like pattern is relatively hypo cellular with elongated rather than plump spindle cells in a densely collagenous background containing scattered lymphocytes, plasma cells and eosinophils. Focal dystrophic calcification and even metastatic ossification can be seen in hyalinised areas⁵.

The myofibroblast was eventually recognized as the principle spindle cell type in tumor. The spindle cells of IMFT are typically uniform and predominantly myofibroblastic in appearance with pale eosinophilic cytoplasm, plump ovoid to tapering vesicular nuclei and one or two small nucleoli. Mild nuclear pleomorphism may be seen, but hyperchromasia is absent. Approximately one half of cases contain scattered ganglion like cells. These are large polygonal cells with abundant amphophilic to eosinophilic cytoplasm, large vesicular nuclei and prominent nucleoli similar to those seen in proliferative fasciitis⁹. Mitotic activity is generally low (0-2 mitosis per 10 HPF), and atypical mitosis are rare^{4,5,9}. Necrosis and vascular invasion have been reported in typical IMT but are very infrequent⁵. Our case did not show any findings of anaplasia or malignancy.

Rarely IMT may undergo histological evolution to a morphologically higher grade lesion with increased cellularity, marked nuclear atypia, frequent mitosis, atypical mitotic figures, and/or necrosis⁵. Myofibroblast are spindle cells having ultra structural features in common with smooth muscle and fibroblast. In IMFT, high cellularity with long plump active myofibroblast with prominent nucleoli can cause confusion with malignancy, in particular with rhabdomyosarcoma. However the lack of atypia, hyperchromasia and abnormal mitotic figure point toward a benign lesion.

IMFT should be diagnosed by routine staining because special stains and immunocytochemistry can be misleading. However, immunohistochemistry shows diffuse reactivity for vimentin and smooth muscle actin.

Rearrangement involving the ALK (anaplastic lymphoma kinase) locus on chromosome 2p23 have been documented in both pulmonary and extrapulmonary IMT providing further support for the neoplastic nature of these lesions and their distinction from other inflammatory pseudotumor¹⁰. IMT are more commonly over diagnosed than under diagnosed.

Tumor resolution or regression has been reported after radiotherapy, chemotherapy and steroid therapy. Local recurrence after incomplete excision is recognized. Few series shows 37% local recurrence, 11% developed distant metastasis and 5 died from disease. This underlies the importance of complete surgical resection whenever possible¹¹.

IV. Figures



Figure 1: Gross photomicrograph revealing a well circumscribed, gray white tumor on serosal aspect of small intestinal segment(ileum).

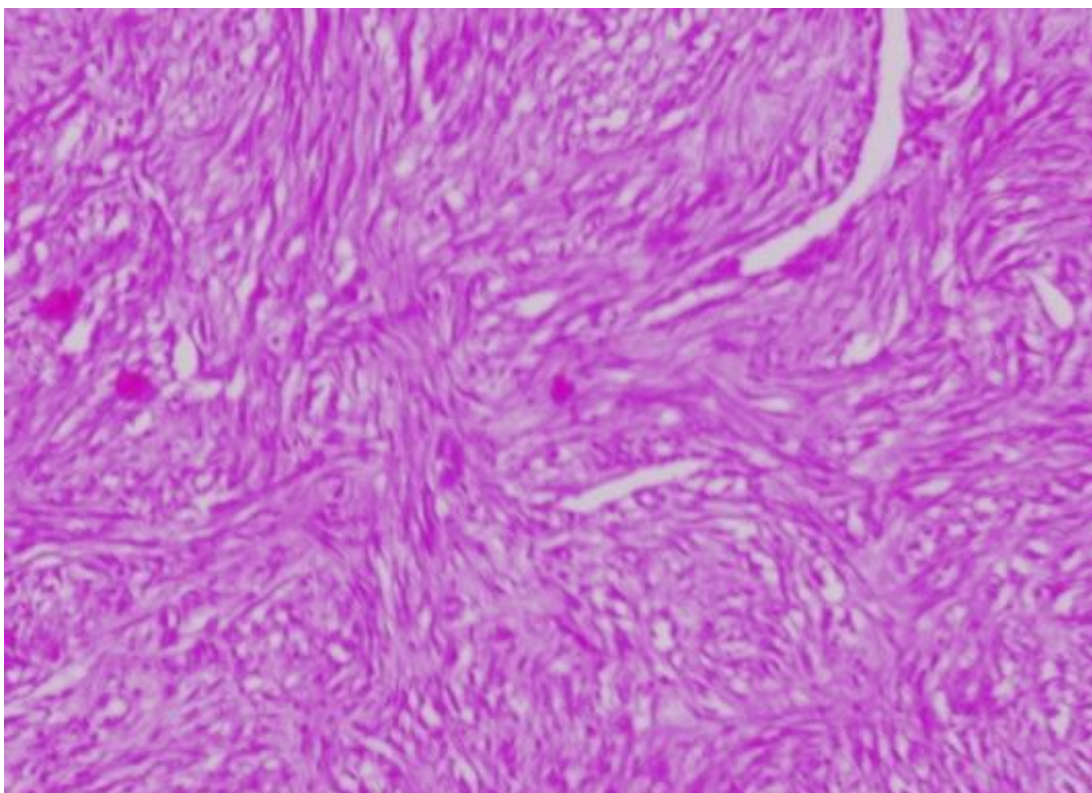


Figure No. 2: Photomicrograph showing spindle cell proliferation in a collagenous matrix(H&E x 10x).

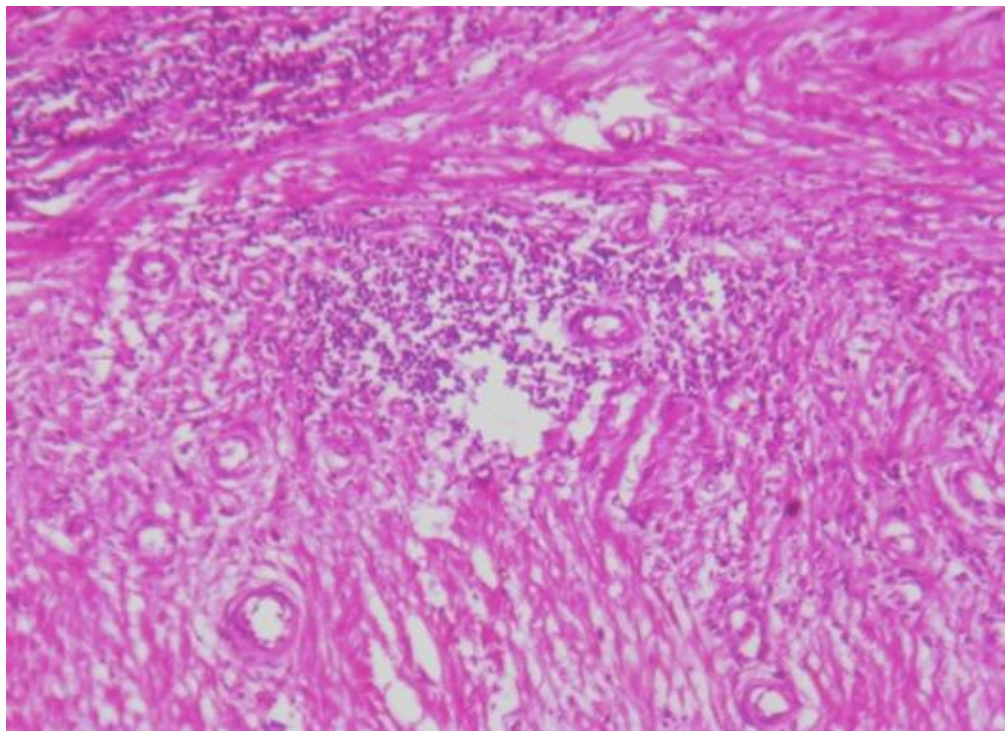


Figure No. 3: Photomicrograph showing inflammatory cell component along with vascular proliferation in a myxoid matrix (H&E x 10x).

V. Conclusion

Inflammatory myofibroblastic tumor is a rare benign tumor and is challenging when it occurs at multiple locations within and outside the abdominal cavity. Due to various clinical presentations, diagnosis can only be made by histological and immunohistochemistry. The local recurrence rate is higher. Thus, in the present case, complete surgical resection was performed to prevent recurrence. This case highlights the histopathological diagnosis of IMFT at unusual multiple sites in a 4 year old child and its confirmation on Immunohistochemistry.

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