Inflammatory Myofibroblastic Tumor of Ileo-Caecal Region
Sahar Iqbal¹, Noshin Wasim Yusuf², Ayesha Imtiaz Malik¹, Ikramul Haq¹, Ishtiaq Ahmed¹

ABSTRACT
Inflammatory myofibroblastic tumor (IMT) is a rare tumor. Other terminologies such as inflammatory pseudotumor, plasma cell granuloma, pseudosarcomatous myofibroblastic proliferation and inflammatory myofibrohistiocyotic proliferation is also used for this tumor. IMT is an uncommon mesenchymal neoplasm which can present at any age & both genders, but usually affect children & young adults. Predominantly it affects the lungs. Besides lung, IMT can also occur in retroperitoneum, mediastinum, liver, spleen & intestines. It has been an issue of great debate for long times, as to whether it is inflammatory reactive process or a neoplastic condition. But now according to recent data, IMTs are considered to be neoplasm of low grade malignant potential. IMT of ileocecal region is a rare tumor sharing histologic features with many other mesenchymal tumors, causing diagnostic dilemma. We report here a case of young female who presented with intestinal obstruction due to a large caecal mass diagnosed clinico-radiologically as carcinoma caecum.

Keywords: Inflammatory myofibroblastic tumor, colon, gastro-intestinal stromal tumor, recurrence, malignant potential.

INTRODUCTION
Inflammatory myofibroblastic tumors (IMTs) is a rare tumor. It was previously known to be non neoplastic, but recent data have indicated its neoplastic nature.¹ Due to its potential for local recurrence (10-25%), persistent local growth and a small risk of distant metastasis (<5%), recent WHO classification tumor grade system classify IMTs as an intermediate grade with very low malignant potential.²³ Predominantly it affects the lung, where it was first described in 1937 as primary lung tumor. Extrapulmonary sites may also be affected, including omentum, retroperitoneum, liver, mesentry & soft tissues.³ GI tract very rarely affected by this tumor. The first case of colorectal IMT was diagnosed in the rectum by Coffin et al in 1995.³⁴ We present one such rare case in the colon, an unusual site of this tumor. A 24 year old female presented with signs & symptoms of peritonitis that is an uncommon presenting feature in gastrointestinal IMTs.

CASE REPORT
A 24 year old female presented to the surgical department, Jinnah hospital, Lahore with history of pain right iliac fossa, nausea, vomiting on & off, and progressive weight loss for one year. The symptoms intensified since 2 days. On clinical examination bowel sounds were absent and signs of peritonitis were present. Peroperatively, a multilobed mass protruding into colon & stretching small intestine with focal gangrenous change were noticed. Right hemicolectomy was done based on these per-operative findings and the specimen was received in Pathology Department, Allama Iqbal Medical College, Lahore, on 2nd October, 2013. A detailed gross examination of the specimen was carried out, which revealed a large polypoidal growth arising from the ileum and protruding into the caecum, completely obstructing the lumen; measuring 18.5 x 14.0 x 9.0 cm, showing firm to hard homogenous appearance on cut section. (Figure. 1 A-C) Representative sections were taken. After routine processing and paraffin block preparation, H&E stained sections were prepared for microscopic examination.

Microscopic examination (Figure: 2) revealed necrotic sloughed mucosal surface with a submucosal cellular tumor, composed of plump spindle shaped cells having pale eosinophilic cytoplasm with ovoid to tapering vesicular nuclei, arranged in a fascicular pattern in

- Mass extending transmurally & give nodular appearance on serosal surface.

Correspondence
Dr Sahar Iqbal, Senior Demonstrator, Department of Pathology, Azra Naheed Medical College, The Superior College Lahore. Cell: 0323-7423670 Email: sahar_moeed@hotmail.com
1 Department of Pathology, Azra Naheed Medical College, The Superior College Lahore
2 Department of Pathology, Allama Iqbal Medical College, Lahore.
colagenous stromal background. The background also showed plasma cell infiltrate along with lymphocytes, forming follicles at places. Sections also revealed evidence of tumor penetration through serosa. However, no marked pleomorphism or mitosis was seen. Histopathological diagnosis of IMT with predominant compact spindle cell pattern was made based on the above mentioned morphological pattern.

**DISCUSSION**

Inflammatory myofibroblastic tumor (IMT), historically also named as plasma cell granuloma, inflammatory pseudotumor, pseudosarcomatous myofibroblastic proliferation. This lesion has evolved overtime, initially being considered a reactive inflammatory lesion to the recent concept of neoplasm of intermediate biological potential, with repeated recurrences and very rare metastatic potential. In contrast to pulmonary IMT, which occurs in mid adulthood, extrapulmonary IMT occurs within the first two decades, and rarely after 30 years of age. IMTs of the GI tract usually present with nonspecific symptoms, such as anorexia, anemia, abdominal pain, and weight loss. Patients with IMTs in the colorectal region usually have the same clinicopathological features as colorectal carcinoma such as altered bowel habits, bleeding per rectum & abdominal mass. Compared to other locations, the prognosis of colorectal IMT is the surgical excision. A range of tumors is strictly monitored. Currently, treatment of choice for the colorectal IMT seems good with less frequent recurrences and low grade malignant potential. Hence there is an utmost need for histopathologists to be aware of this rare but definitely a distinct entity with characteristic clinical, histopathological & molecular features, as complete and well timed surgical excision can be totally curative and helps in avoiding over treatment for a tumor of low grade malignant potential.

**Table 1. Immunohistochemical stains Used for Differential Diagnosis of Colorectal IMT**

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<th>Lesion</th>
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<th>SMA</th>
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<th>ALK</th>
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Most recent data regarding the prognosis of IMT revealed that the anaplastic lymphoma kinase (ALK) gene rearrangements, located on 2p23.2, play a major role in about 50% of cases. IMTs that show nuclear membrane or perinuclear ALK immunoreactivity have more favorable prognosis with low risk for relapse. Anti-ALK treatment with crizotinib can also be offered to such patients, showing good response to this therapy. High PS3 index also considered as bad prognostic factor. No additional data that help in prognosis have been reported till date.

**CONCLUSION**

IMT of ileocecal region is a rare tumor sharing histologic features with many other mesenchymal tumors, becoming diagnostic dilemma. As treatment modalities & prognosis vary so it is vitaly important that distinction should be clearly made among all these cases. Long term follow up of the patient is compulsory considering the high recurrence rate of IMT. Hence there is an utmost need for histopathologists to be aware of this rare but definitely a distinct entity with characteristic clinical, histopathological & molecular features, as complete and well timed surgical excision can be totally curative and helps in avoiding over treatment for a tumor of low grade malignant potential.

**REFERENCES**