CASE REPORT

Inflammatory Myofibroblastic Tumor of Ileo-Caecal Region

Sahar Iqbal[1], Noshin Wasim Yusuf[2], Ayesha Imtiaz Malik[1], Ikramul Haq[1], Ishtiaq Ahmed[1]

ABSTRACT

Inflammatory myofibroblastic tumor (IMT) is a rare tumor. Other terminologies such as inflammatory pseudotumor, plasma cell granuloma, pseudosarcomatousmyofibroblastic proliferation and inflammatory myofibrohistiocytic 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-radiologicaly 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.(2,3) 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.(3,4) 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 2ndOctober, 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.

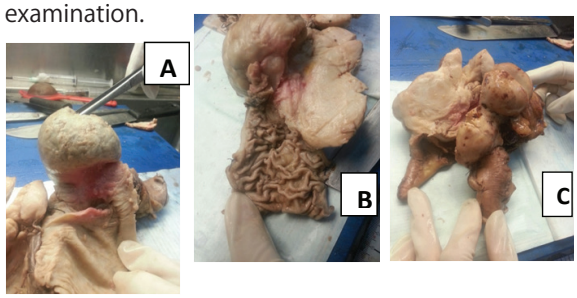

Figure.1: Gross photograph of right hemicolectomy specimen

A) Large polypoidal mass arising from the ileum and protruding into the caecum, completely obstructing the lumen, with ulcerated mucosal surface.
B) Mass measures 18.5 x 14.0 x 9.0 cm, with firm to hard homogenous grey white fleshy appearance on cut surface.
C) Mass extending transmurally & give nodular appearance on serosal surface.

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 plump ovoid to tapering vesicular nuclei, arranged in a fascicular pattern in

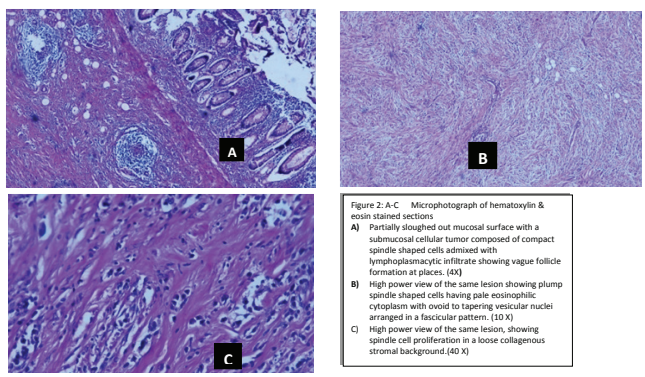

Figure 2: A-C Microphotograph of hematoxylin & eosin stained sections
A) Partially sloughed out mucosal surface with a submucosal cellular tumor composed of compact spindle shaped cells admixed with lymphoplasmacytic infiltrate showing vague follicle formation at places. (4X)
B) High power view of the same lesion showing plump spindle shaped cells having pale eosinophilic cytoplasm with ovoid to tapering vesicular nuclei arranged in a fascicular pattern. (10 X)
C) High power view of the same lesion, showing spindle cell proliferation in a loose collagenous stromal background.(40 X)

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.

collagenous 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, pseudosarcomatousmyofibroblastic 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.[5] 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.[6] 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.[7] Compared to other locations, the prognosis of a colorectal IMT seems good with less frequent recurrences after surgical excision. Malignant behavior of colorectal IMT cannot be totally ruled out, and so long-term follow-up of the patient is mandatory. If the tumor size is larger than 8cm, with aggressive local invasive growth then, this follow up is strictly monitored. Currently, treatment of choice for the colorectal IMT is the surgical excision.[7,8] A range of tumors are included in its differential diagnosis in the GI tract, such as GIST, leiomyoma, schwannoma and spindle cell sarcoma, especially if lesion is necrotic & mitotically active. A panel of immunohistochemistry can be used to sort out the list of differential diagnosis & establish the final diagnosis as shown in table 1.

However, the diagnosis of IMT is not confirmed just on the basis of Immunohistochemistry results. The reason being variablity in expression and lack of specific myofibroblastic markers.

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

	spindle cells IHC spindle cells					
Lesion	**CD 117**	**CD 34**	**SMA**	**Desmin**	**S100**	**ALK**
IMT	_	_	+	+	+	+
Inflammatory fibroid polyp	_	+	+	_	_	_
GIST	+	+	+	_	+	_
SFT	_	+	_	_	_	_
Schwannoma	_	_	_	_	+	_
Leiomyoma	_	_	+	+	_	_
Leimyosarcoma	+	_	+	+	_	_
Fibromatoses	_	_	+	+	_	_

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.9,10 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 P53 index also considered as bad prognostic factor. No additional data that help in prognosis have been reported till date.[7,8,9,10]

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 vitally 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

1. Jain S, Bhargava SK, Upreti L, Mohta L. Inflammatory pseudotumor of sigmoid mesocolon. Indian journal of Radioimaging. 2004; 14:103-4.
2. Kim EY, Lee IK, Lee YS, et al. Inflammatory myofibroblastic tumor in colon. J Korean Surg. 2012; 82:45-49.
3. Coffin CM, Watterson J, Priest JR, Dehner LP.Extrapulmonary inflammatory myofibroblastic tumor (inflammatory pseudotumor). A clinicopathologic and immunohistochemical study of 84 cases. Am J SurgPathol. 1995; 19(8):859–872.
4. Erkan N, Yildiririm M, Yilmaz C, et al. Inflammatory pseudotumor as anunusual case of colonic obstruction: A case report. ActaChirBelg. 2004; 104:462-464.
5. Bjelovic M, Micev M, Spica B, Babic Tamara, Gunjic D, Djuric A et al. Primary inflammatory myofibroblastic tumor of the stomach in an adult woman: a case report and review of the literature. World Journal of Surgical Oncology. 2013; 11(1): 35.
6. Jain A, Kasana S, Ramrakhiani D, Sharma M. Inflammatory myofibroblastic tumor of the stomach in an adult female - report of a rare case and review of the literature. Turk J Gastroenterol. 2012; 23(4): 399-405.
7. Gurzu S, Bara T, Jung L. Inflammatory myofibroblastic tumor of the colon. J Clinical Oncology. 2013; 31(10): e155-e158.
8. Coffin CM, Hornick JL, Fletcher CD. Inflammatory myofibroblastic tumor: comparison of clinicopathologic, histologic, and immunohistochemical features including ALK expression in atypical and aggressive cases. Am J SurgPathol. 2007; 31(4):509–520.
9. Appak YC, Sahin GE, Ayhan S, Taneli C, Kasirga E. Inflammatory Myofibroblastic Tumor of the Colon with an Unusual Presentation of Intestinal Intussusception. Eur J PediatrSurg Rep. 2014; 2: 54-57.
10. Tsuzuki T, Magi-Galluzzi C, Epstein JI. ALK-1 expression in inflammatory myofibroblastic tumor of the urinary bladder. Am J Surg Pathol. 2004; 28: 1609-1614.