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Case Report

Jejunal inflammatory myofibroblastic tumor: a rare entity

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ABSTRACT

Inflammatory myofibroblastic tumor is one of the rare solid tumor occurring in children. Main stay of treatment is surgical resection and in some corticosteroids or NSAIDS may be useful. Here, a case of 3-year-old female with inflammatory myofibroblastic tumor of jejunum is presented, that was evaluated clinically, investigated radiologically and finally histopathology confirmed the diagnosis. No complications occurred at peri and postoperative period. The patient was on regular follow-up and no recurrence had been documented yet in 1 year of follow up. In this article, we reviewed the literature for inflammatory myofibroblastic tumor.

Keywords: Inflammatory fibrosarcoma, Inflammatory myofibroblastic tumor, Jejunum

INTRODUCTION

Inflammatory myofibroblastic tumor is one of the rare solid tumor occurring in children.[1] This term was first given in 1939 by Bunn.[2] This tumor is also known by various terms such as inflammatory pseudo tumor, plasma cell granuloma, inflammatory myofibrohistocytic tumor, inflammatory fibrosarcoma.[3-5] These terms are often confused with soft tissue sarcoma because of its recurrence tendency and local invasion.[5] Main stay of treatment is surgical resection and in some corticosteroids or NSAIDS may be useful.[6]

CASE REPORT

A 3-year-old female child presented with complaints of pain in abdomen off and on for past one month and abdominal distension which her parents noticed one month back. There was no episode of vomiting and constipation. On examination large freely mobile lump, firm in consistency with bosselated surface was palpable in left side of abdomen. Routine blood investigations were within normal limits. CECT abdomen revealed well defined mass lesion of size 10x8x5 cm in left lower abdomen and pelvis with query of ovarian in origin. Patient was taken up for exploratory laparotomy.

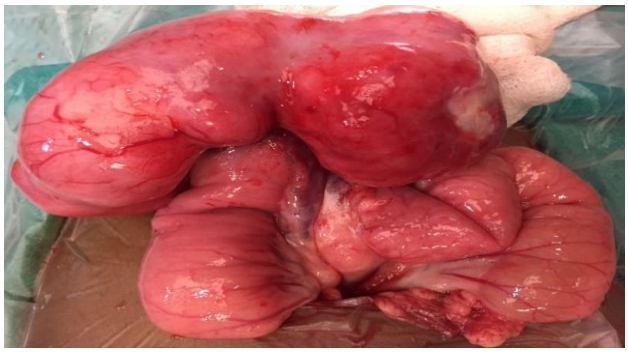

Figure 1: Large bosselated jejuna mass over antimesenteric border.

Large irregularly shaped well encapsulated solid mass with bosselated surface arising from anti mesenteric

border of jejunum, approximately 80 cm distal to duodeno jejunal junction. Mass was resected along with adjacent bowel and end to end jejunojejunal anastamosis was done. Ascitic fluid was negative for malignant cells. Histopatology showed a calcifying inflammatory myofibroblastic tumor. The patient is on regular follow-up and no recurrence had been documented yet in 1 year of follow up.

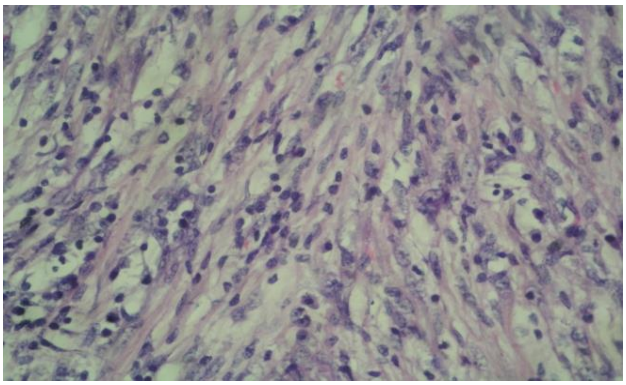

Figure 2: H and E- 20X x 10X- proliferating fibroblasts with lymphocytes and plasma cells.

DISCUSSION

Inflammatory myofibroblastic tumors are solid tumors consisting of spindle shaped cells along with plasma cells, lymphocytes and histocytes.[4,7] This term was first given in 1939 by Bunn.[2] The exact etiology is unknown. According to some authors it is because of immunologic response to an infectious agent and some believe that they are true neoplasm.[1,4] Associated with Epsrein Barr Virus, *Campylobacter jejuni*, previous abdominal surgery has been documented.[8,9]

The most common site documented is lung.[6] Other sites include small and large bowel mesentery, omentum, diaphgram, small and large bowel, appendix, abdominal wall.[6,10] Of all the extrapulmonary sites mesentery and omentum are the most common as stated in study by Coffin and associates. Extrapulmonary tumors are more common in children with mean age of 10 years. More common in females.[6]

Signs and symptoms depend upon the site of tumor. Abdominal mass, abdominal pain, sometimes intestinal obstruction are the common presentation of intra-abdominal tumors.[11] Inflammatory myofibroblastic tumors have variable appearance on CT scan. It has well defined margins, in some calcified and fatty compounds are present.[12] It is sometimes difficult to differentiate from other neoplasm as there is high uptake of trace in PET scan.[13]

The rate of recurrence of these tumors following surface is 18-44% as stated by Coffin and associates in their study.[14] Because of local recurrence tendency and mild risk of metastasis WHO has classified these as tumor of intermediate biological potential.[15] Common differential diagnosis include GIST, fibromatosis, solitary fibrous tumor, leiomyosarcoma.[16]

Tumor is composed of chronic inflammatory cells including lymphocytes, plasma cells, sometimes histocytes along with spindle cells. Stroma can have focal areas and calcification. There is variable proportion of inflammatory cells to spindle cells. Different types of patterns are identified microscopically.[6]

- Fascities pattern: spindle cells in myxoid stroma with inflammatory cells.
- Proliferating pattern: in this mitotic figures are seen.
- Sclerosing pattern: having focal areas of calcification.

Rarely profound cellular atypia can be seen in tumors. Poor prognosis is associated with increased mitosis and cellularity.[6] Coffin et al in their study found mitosis ranging from 0-2 per 50 HPF is associated with favorable prognosis as compared to mitosis ranging from 1-7/50 HPF with unfavourable prognosis. They also suggested IMT to be benign, non metastatizing tumor having tendency foe recurrence.[6]

On cut section of tumor there is nodular and lobular appearance with myxoid background and few area of haemorrhage and necrosis. Literature reports the mean tumor size of 8cm.[17]

Myofibroblastic nodules of spindle cells is supported on immunostaining which is positive for smooth muscle actin, vimentin is about 50% cases of IMT ALK is positive. ALK given is located on chromosome 2P 23 and its positivity is associated with favorable outcome. Spindle cells ultrastructural study revealed common features as seen in smooth muscle cells and fibroblasts.[14]

Birelli and coworker in their study found that around 50% of extrapulmonary IMFT in pediatric age group are aneuploid.[18] Meis and Enzinger in their study of 38 cases found to have 3 patients with distant metastasis and 8 had local invasion.[19]

Reports are there in literature supporting the neoplastic nature of the tumor. Surgical resection of tumor is mainstay of therapy, and in some cases corticosteroids and NSAIDS are useful. For local recurrence, also re-excision is treatment of choice.[6] Tothova et al in their study recommended radical surgery as main stay owing to high rate of recurrence.[20] There is local recurrence of around 23% following complete excision and rarely these undergo malignant transformation. Risk of distant metastasis is <5% and 5-year survival rate is 87%.[21]

Murga-Zamalloa et al studied the role of ALK inhibitor, Crizotinib, and its trials have shown good results.[22] Crizotinib was also recommended in the management of IMFT in the study by Tothova et al. It had further lead to

the development of more selective ALK inhibitors which were useful in cases of crizotinib resistance mutations.[20] Corticosteroids, NSAID's and radiotherapy have shown good results in ALK negative IMFT were surgery in not feasible. Corticosteroid monotherapy was recommended by Carswel et al for rapid resolution of disease.[23]

CONCLUSION

Intra-abdominal inflammatory myofibroblastic tumors is one of the rare entity. Accurate diagnosis should be made as it associated with rate of recurrence. Long term follow up of patient is necessary. Complete surgical resection is treatment of choice.

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