ORIGINAL RESEARCH PAPER
Volume - 11 | Issue - 07 | July - 2022 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

INFLAMMATORY MYOFIBROBLATIC TUMOR OF TERMINAL ILEUM : A REPORT OF 2 CASES

Pathology

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ABSTRACT

Inflammatory myofibroblastic tumor (IMT) is a rare but benign entity masquerading as a malignant tumor. The tumor most commonly occurs in the pulmonary system of children and young adult, although it may rarely develop in older patients and other organs. Symptoms are non-specific and depend on the location of the tumor. The gastrointestinal tract is rarely the primary site of origin for this lesion.We report two cases of IMT of terminal ileum; one in a 5 month old child while another in a 61 year old female.

KEYWORDS

Inflammatory myofibroblastic proliferation, terminal ileum

INTRODUCTION

Inflammatory myofibroblastic tumor (IMT) is a kind of mesenchymal tumor characterized with proliferation of myofibroblast spindle cells and prominent infiltration of plasma cellsand or lymphocytes. IMT is rare and its true incidence and prevalence remains unclear [1].The lesion, as primarily described by Bahadori and Liebow in 1973, is a reactive/inflammatory process in the pulmonary system mostly occurring in children and young then, many cases have been reported in older patients in addition to extra-pulmonary sites[2]. Here we report two cases of IMT of terminal ileum in a 5 month old infant and a 61 year old adult .

CASE REPORTS

CASE 1:

5 months old infant presented with fever and loose stools. There had been no evident congenital abnormalities at birth, and he had no family history of disease. On a physical examination he had a palpable mass in the right lower abdomen. Xray was consistent with persistently dilated bowel loops (transverse colon). A clinical diagnosis of duplication cyst was kept was taken to operating room for the removal of the mass. Operative findings showed a mass consisting of solid tissue in the terminal ileum arising from the mesentry and was separated easily and sent for the histopathology examination.

We received a single grey white to grey brown soft tissue piece measuring 5x4x3 cm. Outer surface was encapsulated and shiny. Cut section of the mass showed a well circumscribed grey white glistening tumor with no areas of necrosis and hemorrhage. On microscopy the lesion was comprised of myofibroblastic and fibroblastic spindle cells with a background of inflammatory infiltrate comprising of lymphocytes, plasma cells , histiocytes and eosinophils and abundant blood vessels and vascular channels. The spindle cells have slender to plump bland vesicular nuclei with fine chromatin, small conspicous nucleoli and eosinophilic cytoplasm. Foci of dense hyalinized stroma and fibroblastic stroma was also noted. Scattered macrophages with hemosiderin pigment and focal myxoid areas were also seen. A diagnosis of inflammatory myofibroblastic tumor of distal ileum was given.

Case 2:

A 61 year old female presented with pain abdomen and abdominal swelling for a duration of four month which increased with time. Patient had undergone ventral wall hernial repair 7 months back. On examination, a swelling of approximately 13x14 cm in size was seen in the paraumblical region with umblicus pushed left laterally. Presence of scar with no dilated veins or visible peristalsis. Cough impulse was present. On auscultation no bowel sounds were present. On CT abdomen, impression of recurrent incisional hernia with herniation of bowel loops was given. Patient then went into surgery for hernia repair. Operative findings showed a defect of 20x14 cm present in the omentum which was repaired with a mesh. There was also an incidental nodule found over the antimesentric border of the terminal ileum which was resected out and was sent for the histopathological examination.

We received a single grey white to grey brown soft tissue piece measuring 1.2x1.2x 0.8 cm . the cut section was grey white to grey brown to tan brown. Microscopy revealed intestinal tissue fragment showing ileal mucosa. Serosal surface shows a cellular lesion composed bland-looking spindle cells (myofibroblasts) with a collagenous and myxoid background accompanied by various diffusely scattered inflammatory cells, including plasma cells, lymphocytes and eosinophils and occasional mast cells. No mitosis, necrosis or hemorrhage was present and hence a diagnosis of inflammatory myofibroblastic tumor was given.

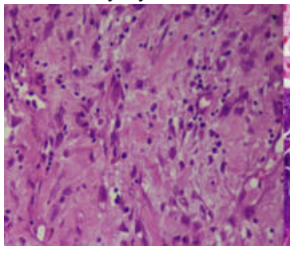

Figure 1: (Case 1) High power view of the tumor composed of myofibroblastic and fibroblastic spindle cells in a background of lymphocytes, plasma cells and eosinophils. (H&E.400X)

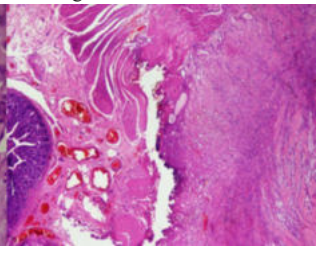

Figure 2: (Case 2) Low power view of ileal nodule revealing ileal mucosa with muscularis propia bundles and serosal surface showing collagenous bands with proliferation of myofibroblasts and inflammatory cells(H&E,100X)

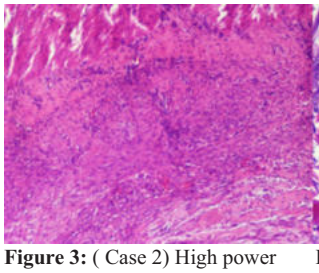

Figure 3: (Case 2) High power view of the ileal nodule showing muscularis propria bundles along with myofibroblastic and fibroblastic spindle cells poliferation (H&E,400x)

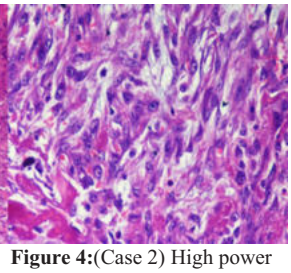

Figure 4:(Case 2) High power view revealing plump spindled cells with vesicular nuclei, fine chromatin , conspicous nucleoli and eosinophilic cytoplasm along with mast cells, eosinophils in a myxoid stroma (H&E 400X)

DISCUSSION

IMT is defined as “proliferation of fibroblasts and myofibroblasts, infiltrating inflammatory cells” and commonly occurs in the young people [3]. The tumor has been reported to occur in the systemic organs and soft tissues throughout the body, after originally being described in the lung; however, intestinal IMT, especially in the ileum, is extremely rare.

They are not uncommon and typically seen in children between the

ages of 2 years and 16 years[4]. Infantile cases have also been described [5].The presenting symptoms and signs depend on the location and growth pattern of the primary lesion. These patients often present with abdominal pain and fullness, fever, palpable mass, weight loss, and other vague nonspecific symptoms. Obstruction or intussusception is the most frequent initial symptom when an IMT is located in the small bowel [6].

Pathogenesis of IMTs are unknown but development of IMT has been described after trauma, surgery or infection [7] as seen in our case 2 in which IMT developed after hernia repair surgery. There have also been some reports regarding the association of IMT with Behcet's disease, Hodgkin's disease or chronic peptic ulcer [8].

Clinically and radiologically it may often mimic a malignant tumor. CT and MRI shows findings of obstruction. Since IMTs have no distinctive radiologic and clinical findings, histologic confirmation is necessary in all cases to exclude malignancy.

Gross appearance of IMTs are usually well circumscribed solid mass. May be present as multiple nodules depending on the anatomic site. Histopathological findings in typical IMT are a fasciitis like compact spindle cell proliferation with areas of myxoid change and hypocellularity showing a collagenous back ground [9]. Various numbers of mixed inflammatory cells including plasma cells, lymphocytes, eosinophils and rarely foamy macrophages are seen. Typically the spindle cells of the IMT express vimentin, smooth muscle actin and other markers which correspond to the myofibroblastic nature of these cells[3]. IMT shows three major subtypes in histologic appearance: fibromyxoid and vascular pattern, proliferating pattern, and sclerosing patterns. Sclerosing pattern was seen in our case 2.

Differential diagnosis includes a spindle-cell lesion from mesenchymal neoplasms or other malignant masses in the abdomen including leiomyosarcoma, inflammatory fibrosarcoma, idiopathic sclerosing mesenteritis, spindle-cell carcinoid rhabdomyosarcoma, and malignant fibrous histiocytoma.

Because of the rarity of this proliferation in the terminal ileum, its natural history and biological potential are still uncertain. IMTs generally have a benign course, and recurrence or distant metastasis is rare. The incidence of local recurrence has been reported to be 15–37 % when an IMT is located in the mesentery or the retroperitoneum[10]. Surgery is the main stay of treatment and patient should be kept under follow up. IMTs are locally aggressive lesions, and Alag gio et al. [11] reported that the local recurrence rate of IMTs in children was 23%.

CONCLUSION

IMT of the small intestine is a very rare but locally aggressive lesion and recurrence. Itshould be considered in the list of the differential diagnoses of spindle cell tumors in any part of the body of patients at any age, particularly children and young adults.A precise diagnosis should be based on histological findings, and complete surgical resection is necessary. Clinical and laboratory follow-up is mandatory because of an increased risk for local recurrence.

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