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Inflammatory myofibroblastic tumor of ileum in adult female patient presenting as ileocolic intussusception

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Abstract

Inflammatory Myofibroblastic Tumors are classified among very rare but proven tumors of the body that have been surgically removed. They are made up of myofibroblastic spindle cells and they usually arise in mucosal surfaces and mesentery. A lot of terms have been applied to this lesion which includes, plasma cell granuloma, inflammatory pseudotumor, pseudosarcoma, fibrous xanthoma, lymphoid hamartoma, myxoid hamartoma, inflammatory myofibrohistiocytic proliferation, benign myofibroblatoma, and lately, inflammatory myofibroblastic tumor. Due to its rarity, no standard treatment protocol has been established so far and it is only understood to have complete surgical resection of the tumor followed by surveillance for high risk for recurrence. In this case report, we discuss a case of a young female patient who suffered with upper abdominal pain for three months which was shadowed by wrong diagnosis and wrong intervention but later discovered with ileocolic intussusception and histopathology report of tumor revealed IMT of ileum. Which is why it's strongly recommended through this case report to implement further studies to have a better and definitive approach towards the management and treatment of this disease.

Keywords

inflammatory myofibroblastic tumors; ileum; ileocolic intussusception; abdominal pain

Abbreviation

IMT: Inflammatory myofibroblastic tumors; CT-scan: Computed tomography scan

Introduction

According to statistics done in a study, IMT develops at a mean age of 9.7 years, and in 36 of 84 cases (43%), IMTs arose from the mesentery and omentum and only 1 case (1.2%) exhibited an IMT of ileal origin [1]. It is usually benign but also has malignant type and is accompanied by various clinical manifestations because of its diversity for its site of origin. It can affect both genders and doesn't have a

genetic predisposition. Due to rarity of this disease, the pathogenesis, risk factors, clinical manifestations and outcome are not well known. Data is very scarce, does not describe natural history of disease after resection and what adjuvant treatment should be offered after complete resection. However, it was first discovered by Bunn in 1939 in lungs and later on various other extra pulmonary sites were described [2]. Commonly reported areas of origin are lungs, orbit, peritoneum including lining of the abdominal cavity and internal organs and mesentery [3]. Most reported patients are asymptomatic while others have symptoms relevant with the location of tumor like cough, fever, dyspnea and abdominal pain. Weight loss and anorexia are uncommon. It usually occurs in children and young adults but it can develop in any age. Common hypothesis suggests an allergic or auto-immune mechanism or infectious origin, that's why named so. Due to its non-specific symptoms, preoperative diagnosis of IMT is very difficult. However, CT-scan can be used to look for the cause of persistent pain or other signs of gastrointestinal upset. The treatment of choice is complete resection. It is both diagnostic and therapeutic and in the end, histopathology report is the necessary tool for confirmation of this disease. Resection should be complete with negative margins because an incomplete resection increases the risk of recurrence. Chemotherapy is only used in cases of multifocal, malignant invasive lesions or in cases of local recurrence.

Case Report

A 28 year old married nulliparous female with no previous known co-morbids presented in general surgery OPD with complaint of right hypochondrium pain for three months. Initially it was mild in intensity associated with nausea, normal bowel habits and not related with food intake. But for last one month, her pain had increased in intensity. It used to become severe and generalized in abdomen after taking meals associated with feeling of not being able to pass flatus. She would suffer from constipation for 3-4 days followed by diarrhea for next 1-2 days. Due to persistent pain often accompanied with vomiting, patient had decreased her oral intake and lost about 3kgs weight. Primarily she went to a small surgery clinic in her hometown and got USG abdomen and pelvis which showed right ovarian cyst. Hence under doctor's recommendation at the time she underwent right ovarian cystectomy to get relief from the pain. But even after surgery, patient's symptoms did not resolve so she had a CT-Scan Abdomen with IV contrast which showed intussusception complex with intraluminal mass of soft-tissue density, dilated loops of small bowel proximal to the tissue mass, hepatic focal lesion and few enlarged lymph nodes in para aortic and mesenteric regions.

Three days earlier to presentation, she began to experience an intensified and generalized abdominal pain associated with nausea, vomiting and history of decreased gas passing during this period. So she came to tertiary care hospital for a second expert opinion. On examination, she was vitally stable and not in acute distress. The abdomen was faintly distended with mild generalized tenderness on palpation. Bowel sounds were slightly exaggerated. Rectal examination was normal. All her baseline lab workup was normal including serum electrolytes. She was admitted and decision for urgent surgical intervention was made.

She underwent exploratory laparotomy and intraoperatively, ileocolic intussusception was discovered with tumor as leading point 15cm from ileoceacal junction (Figure 1). It was about 4cm x 4cm

x 4cm in size and regular in shape completely obstructing the lumen and distal bowel collapsed (Figure 2). Liver was completely normal. Right hemicolectomy was done with side to side functional end anastomosis. Postoperative period was uneventful. Patient remained vitally and clinically stable; no adjuvant therapy was employed and she was discharged home on 4th post-operative day.

Histopathology specimen revealed Inflammatory Myofibroblastic Tumor of ileum with negative proximal and distal margins. Cut surface showed tan gray, homogeneous mass. Microscopically, submucosa comprised of spindle shaped cells, background tissue is myxoid and contains abundant mixed inflammatory infiltrates comprising of lymphocytes, plasma cells, eosinophils and mast cells. No necrosis or atypia present. Five lymph nodes recovered showing reactive changes. Immunohistochemistry was negative for ASMA, DOG1, ALK, CD117 and CK7.

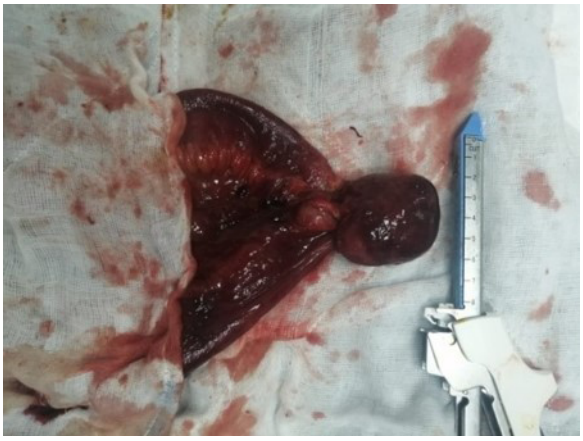

Figure 1: Tumor of Ileum 15cm from ileoceacal junction.

Figure 2: Cut section of Ileum Tumor

Discussion

The Literature review for this disease is mostly descriptive and it only reflects uncertainty regarding true biologic nature of these lesions. Lymphocytes, plasma cells, histiocytes, fibroblasts and myofibroblasts are so far the basic components of IMT, present in inconstant proportions.

Recently, the concept of IMT being a benign reactive lesion has been challenged because of documented evidence of recurrences as high as 37% with the presence of regional metastases and cytogenetic evidence of acquired clonal chromosomal abnormality [4]. However, the issue of reactive versus neoplastic pathogenesis of this lesion remains unsolved and needs further work. The malignant nature of this tumor is mirrored by some cytogenetic and immunofluorescence anomalies, mainly chromosome 2p23 involving the Anaplastic Lymphoma Tyrosine Kinase (ALK) receptor and its fusion with clathrin heavy chains. ALK-1 expression is highly specific for IMT, but it is not 100% sensitive, depending to some extent on the site of origin. ALK-1 negative IMTs are morphologically indistinguishable from ALK-1 positive cases. And although ALK-1 plays an important role but no clinical, morphological or prognostic difference is found associated with ALK-1 status of the IMT [5].

Intussusception is a disease more common for pediatric age group. It is rare and difficult to diagnose in adults because patients usually present with non-specific, vague symptoms like abdominal pain, nausea and vomiting which mimic a lot of other more common causes of acute abdomen. Latest studies show that

approximately 30% of all small bowel intussusceptions are caused by malignancy, whereas the rest of 60% are caused by benign lesions and 10% are idiopathic [1].

All in all, IMT if resected with negative margins usually follows benign course with favorable outcomes. But all the patients diagnosed with IMT should undergo regular surveillance for recurrence. In our case, we plan to follow the patient with six monthly CT-scans for two years and then yearly follow up to five years postoperative period.

Intussusception itself is a very rare presentation in adults and having Inflammatory Myofibroblastic Tumor as its cause makes it more unique and noteworthy. Till this day, there is still lack of standard treatment regime and deficiency of varying data makes it difficult to have a solid management plan. Hence, it is strongly recommended through this case report to have more studies done on rare diseases like Inflammatory Myofibroblastic Tumor so that patients can have a more certain and promising future regarding their disease and prognosis.

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