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Case Report

Sclerosing Mesenteritis Masquerading as Ileal Malignancy

Vibhu Jain[1], Santhosh Irrinki[1], Amanjit Bal[2] and Kailash Chand Kurdia[1,3]*

[1]*Department of General Surgery, Post Graduate Institute of Medical Education and Research, India*

[2]*Department of Histopathology, Post Graduate Institute of Medical Education and Research, India*

[3]*Department of GI Surgery and Liver Transplantation, All India Institute of Medical Sciences, India*

Abstract

Sclerosing mesenteritis is a rare non-neoplastic entity associated with chronic inflammation of the mesentery. The exact cause remains unknown; however, postulated causes include autoimmunity, abdominal trauma and abdominal infections among others. Clinical features of the disease include abdominal pain, vomiting, fever of unknown origin, or even intestinal obstruction. The natural course of the disease can vary between being self-limited or requiring medical and/or surgical intervention. Here we present the case of a patient with complaints of vague abdominal pain and nausea and an abdominal lump in the right iliac fossa. CECT abdomen was suggestive of a mass in close relation to ileal loops, and PET/CT showed an FDG avid lesion in the right iliac fossa close to the terminal ileal loops. A laparotomy was performed with resection of the mesentery and involved bowel segments. The final histopathological diagnosis however came out to be consistent with a benign entity of sclerosing mesenteritis.

Keywords: Sclerosing mesenteritis; Mesentery; Ileal malignancy; GIST

Introduction

Sclerosing mesenteritis encompasses a spectrum of diseases involving fibrosis and inflammatory changes in the bowel mesentery, commonly presenting in the 5th or 6th decade [1]. While majority of the patients are asymptomatic, common clinical features include abdominal pain, distention, nausea, vomiting, and weight loss [2].

In the case presented here, the clinical, radiological, and histopathological examinations were indicative of a malignant lesion of the right iliac fossa, most likely originating from the terminal ileal loops, but the lesion ultimately turned out to be a benign entity-sclerosing mesenteritis.

Case Presentation

A male patient in his 50s without any comorbidity presented with dull aching abdominal pain for 1 month and low-grade fever for 10 days. On examination, there was a firm intraperitoneal 6x6cm mass in the Right Iliac Fossa (RIF) and peri umbilical region with ill-defined borders.

The blood workup was grossly normal. On CECT abdomen there was a 5.5 cm × 4.7 cm × 4.6 cm lobulated, heterogenous lesions in RIF with origin from the terminal ileum, likely Gastrointestinal Stromal Tumor (GIST). An ultrasound-guided biopsy from the lesion was suggestive of a spindle cell neoplasm. PET scan showed an FDG avid 4.7 cm × 4.2 cm × 5.8 cm lesion in the right iliac fossa close to the distal ileum (Standardized Uptake Value (SUV) maximum of 5.8) and a 1.5 cm × 1.4 cm lesion in the left lung apex (SUV max 68) (Figure 1). PET-guided FNAC was planned for the pulmonary nodule but could not be done owing to its difficult location and small size.

Based on the clinical, radiological and histopathological investigations, an ileal malignancy, likely GIST was suspected and the patient was planned for a laparotomy. Intra-operatively, a mass in the distal ileal mesentery densely adhered to the terminal ileum, caecum and ascending colon, D2-D3 junction and right ureter causing upstream ureteric dilatation was seen. A right hemicolectomy with ileocolic anastomosis, sleeve duodenal resection, and feeding jejunostomy was performed. The ureter could be separated from the mass after meticulous dissection. The cut section of the bowel showed normal mucosa (Figure 2). The postoperative course was grossly uneventful and the patient was discharged on postoperative day 9.

The final histopathological report was that of a lesion with fibroblastic proliferation, with underlying hyalinised stroma and infiltration of plasma cells and lymphocytes, and the absence of any neoplastic cells. IgG4 Immunohistochemistry (IHC) highlighted a few plasma cells, however, the number was inadequate to qualify as IgG4-related disease, and IHC for SMA highlighted proliferating fibroblasts and myofibroblasts only, and overall features were suggestive of sclerosing mesenteritis (Figure 3). At one year of follow-up, the patient is doing well with no clinical or radiological signs of recurrence.

Discussion

Sclerosing mesenteritis is a rare entity with a prevalence of less than one per cent, being twice more common in males than in females. It was first reported by Jura in 1924, who called it retractile mesenteritis [3]. Currently, sclerosing mesenteritis is an umbrella term encompassing mesenteric panniculitis, retractile mesenteritis and mesenteric lipodystrophy.

The etiopathogenesis remains largely unknown, and the

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***Corresponding author:** Kailash Chand Kurdia, Department of GI Surgery and Liver Transplantation, All India Institute of Medical Sciences, New Delhi, 110029, India, Tel: +91-6394358323

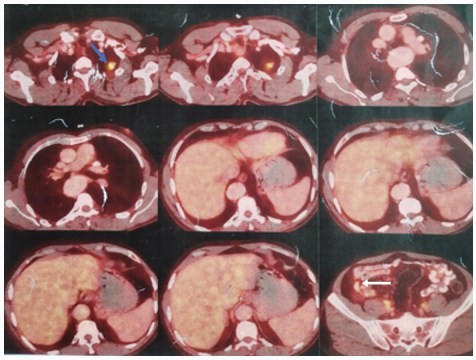

Figure 1: PET scan images showing the FDG avid pulmonary nodule (blue arrow) and right iliac fossa mass in close relation with terminal ileal loops (white arrow).

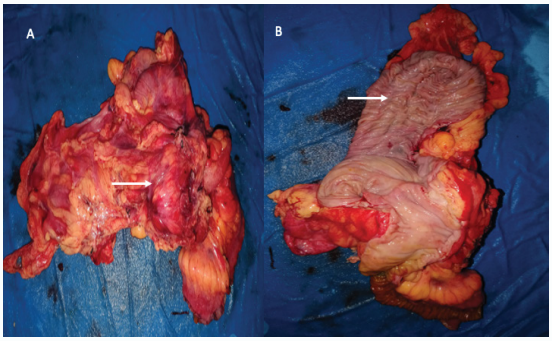

Figure 2: a) Resected right hemicolectomy specimen with the mesenteric mass (arrow); b) Cut section of bowel showing normal mucosa (Arrow).

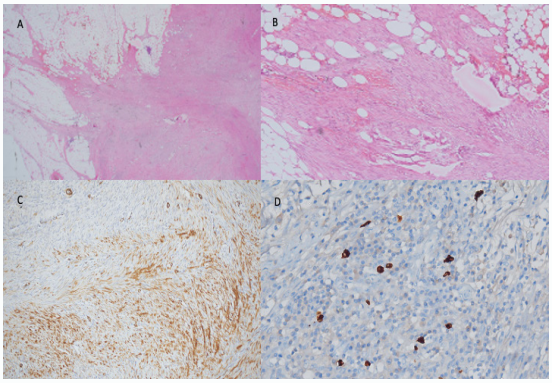

Figure 3: Histopathological images (a) Proliferation of fibroblasts in mesenteric fat x100 (b) High power of fibroblasts x 200 (c) SMA positivity in myofibroblasts (d) IgG4 staining of plasma cells, though insufficient in number to qualify as IgG4 related disease.

proposed etiological factors include prior abdominal surgery/trauma, autoimmunity, paraneoplastic manifestation of a malignant process, and infectious agents including tuberculosis and even secondary to IgG4-related disease [4]. In our case, though IgG4-positive plasma cells were seen in the resected specimen, however, their number was insufficient to label it as an IgG4-mediated disease and the etiology remains idiopathic in this case.

The most common presenting symptoms of SM include abdominal pain, bloating and distention, diarrhoea, and weight loss [5]. Our patient also had a similar presentation, and retrospectively we could attribute the fever to the disease perse.

The most common neoplastic mimics of SM are lymphoma, carcinoid, desmoid tumors and GIST [6]. In our case, radiologically, the lesion was thought to be arising from the bowel loops, and keeping the FDG avidity in mind, the suspicion of a malignant tumor was high.

The role of imaging for diagnostic purposes remains controversial. Tumor pseudo capsule and fat ring sign can be seen on axial CT scan [7]. However, these signs are not very sensitive or specific, and neither of them could be appreciated in our case. PET scan in our case did not help us in reaching a definitive diagnosis. In a study by Akram et al, the diagnosis was established at laparotomy with biopsy in 65%, laparoscopy with biopsy in 25%, and CT-guided biopsy in 10% of cases [5].

Pathologically, SM involves inflammation, fibrosis and fat necrosis of mesentery in varying proportions. Histological specimens with more fibrosis are categorized as retractile disease, as can be seen in this case, fat necrosis with an inflammatory component is panniculitis, and predominately fat necrosis is lipodystrophy [8]. Treatment options include the use of anti-inflammatory drugs, tamoxifen and steroids, either alone or in combination [1]. Indications of surgery include symptomatic presentation, features of small bowel obstruction, or diagnostic dilemma, as was seen in this case.

Owing to the rarity of this disease, there is a paucity of data pertaining to the demography, natural course and treatment options for the disease. We present this case report hoping it adds to the literature and aids in a better understanding of the disease.

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