Abdominal Pain: The Double Trouble!!!

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Background	Mesenteric cystic masses are an uncommon entity seen in surgical practice. They usually occur as single lesion and an independent entity. Surgical excision in such scenarios is often curative. However, preoperatively, they may pose a diagnostic dilemma and may mimic other mesenteric masses, like lymph node cavitation or cystic mesenteric metastasis. There may also be rarely an association with another disease pathology.
Summary	We herein report an unusual case of a young adult gentleman who has multiple cystic lymphangioma scattered throughout the small bowel mesentery in the background of celiac disease and associated with hemolytic anemia and autoimmune hepatitis. The patient had previously been prescribed empirical antitubercular therapy without any symptom resolution. The clinical approach and management is detailed here in this case report.
Conclusion	Cystic mesenteric masses are rare and a detailed and appropriate work up is needed before starting a patient on empirical antitubercular treatment especially in endemic countries.
Keywords	mesenteric lymphangioma; celiac disease; abdominal tuberculosis; lymph node cavitation

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Case Description

Cystic mesenteric masses are infrequently seen in surgical practice and common differentials include mesenteric lymphangioma, lymph node cavitation (as in tuberculosis, celiac disease, or necrotic abdominal metastasis), peritoneal hydatidosis and ruptured cystic neoplasms.[1] We hereby report an unusual case of a young male with multiple mesenteric lymphangioma in the setting of an underlying celiac disease.

A thirty-year-old gentleman presented to the surgical outpatient department with complaints of recurrent central abdominal pain for four years. The patient had been evaluated at another primary healthcare facility with an abdominal ultrasonography (USG), which revealed multiple enlarged mesenteric lymph nodes without any bowel wall thickening, on the basis of which he was given empirical antitubercular treatment (ATT) twice for six months each. Due to persisting symptoms, he then presented to our hospital where he was found to have pallor, body mass index (BMI) of 18.5kg/m^2 and multiple intraperitoneal mobile nodular nontender lumps in the left side of his abdomen. On investigation, he had microcytic hypochromic anemia without any hemolysis on peripheral smear. Further evaluation depicted low Serum Ferritin, normal hemoglobin electrophoresis and a positive direct Coomb's test. His liver function test revealed raised serum transaminases without hyperbilirubinemia. USG of the abdomen revealed multiple cystic masses without internal vascularity, with some showing debris within. Abdominal magnetic resonance imaging (MRI) (Figure 1) demonstrated multiple fluid signal intensity cystic lesions scattered in the small bowel mesentery, especially in the left hypochondrium and lumbar region.

Some of the lesions showed heterogeneous signal on T2 Weighted images and hyper-intense wall on T1 weighted sequence, suggesting hemorrhage within. Possibility of multiple mesenteric veno-lymphatic malformations was kept. Also, the visualized vertebrae appeared to have heterogeneous signal, and spine screening suggested bone marrow re-conversion (Figure 2). With such hematological and imaging features, an upper gastrointestinal endoscopy (UGIE) and anti-TTG antibodies were done to rule out associated celiac disease. His serum anti-TTG was elevated. UGIE showed scalloping of duodenal mucosal folds and duodenal biopsy suggested celiac disease.

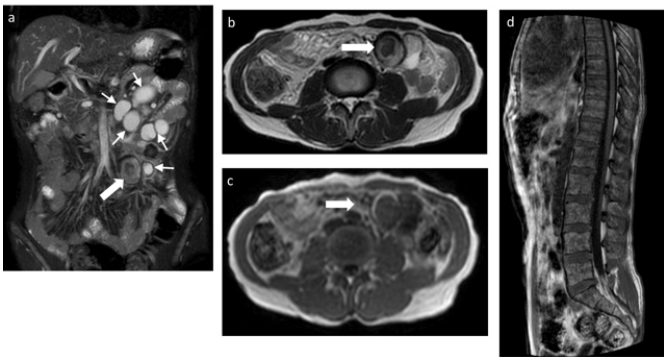

Figure 1. A: Coronal Short tau inversion recovery (STIR) image shows multiple fluid signal intensity (T2 hyperintense) cysts in the mesentery in the left hypochondrium and lumbar region (arrows), showing conglomeration. Few of the cystic lesions were mixed signal intensity on STIR (solid arrow). **B:** Axial T2-weighted (T2W) MR image shows heterogeneous signal with hypointense rim, suggesting haemorrhage within the cystic lesion. **C:** Axial T1W MR image depicts hyperintense periphery, indicative of hemosiderin. **D:** Sagittal T1W MR image depicts heterogeneity within the vertebral marrow with multiple tiny ill-defined foci of mild T1 hypointensity, suggesting marrow reconversion.

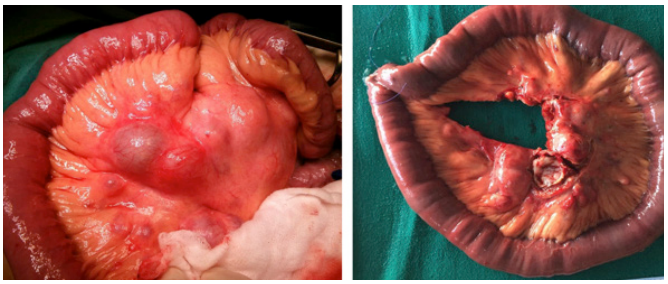

Figure 2. Intraoperative image showing multiple cystic lesions in the small bowel mesentery.

The patient underwent exploratory laparotomy wherein large congruent multiple cystic masses in the mesentery of jejunum and ileum were present for around 100 cm, with few similar small scattered lesions in other parts of small bowel mesentery. Resection of the larger congruent masses with the associated bowel segment was done, trying to take out as many lesions as possible without compromising the remaining bowel vascularity. The ones left behind were either deroofed and marsupialized or left undisturbed in an attempt to salvage as much bowel length as possible. Patient had an uneventful postoperative recovery. The histopathological examination revealed multiple mesenteric cysts with ileal and jejunal segments showing blunting of villi with increased intraepithelial lymphocytes and crypt hyperplasia (Figure 3). The histology of liver biopsy (done in view of raised enzymes) suggested autoimmune hepatitis.

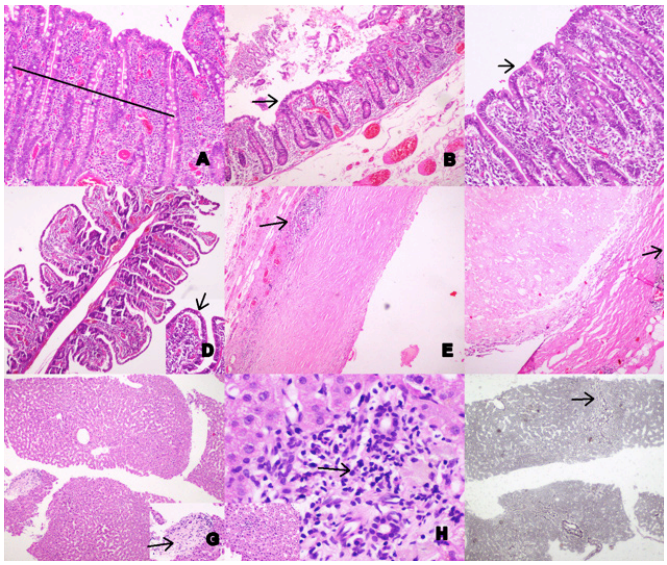

Figure 3. Photomicrograph shows duodenal biopsy with crypt to villous ratio 1:1, crypt hypeplasia and increased intra-epithelial lymphocytes **A:** (x100). Sections from jejunum also shows a C:V ratio of 1:1 **B:** (x40), with increased IELs (arrow) **C:** (x100). Sections from ileum also showed increased IELs (arrow, inset), though the C:V ratio was maintained **D:** (x40). Sections from mesenteric cysts showed thick fibrocollagenous cyst wall with aggregates of lymphoid cells (arrow) **E and F:** (x 40). Intraluminal proteinaceous fluidy material can be appreciated **F:** (x40). Liver core biopsy showed maintained architecture, however the portal tracts were expanded and showed fibrosis (arrow, inset) **G:** (x40). Interphase hepatitis and lymphocytic bile duct injury was noted (arrow) **H:** (x200). Silver reticulin stain showed extension of short fibrous septae within the hepatic lobules (arrow) I: (x40).

Based on the above examinations, a final diagnosis of multiple mesenteric lymphangioma with celiac disease was made. The patient is presently on gluten free diet (GFD) and is symptomatically better at ten months follow-up. A review MRI abdomen was done, which did not show any progression of remaining mesenteric lesions.

Discussion

Cystic lesions of the mesentery are uncommon and their differentials include mesenteric lymphangioma and mesenteric lymph node cavitation. Mesenteric lymphangiomas account for <1 percent of these lesions.[2] They are most commonly congenital but may be seen in young adults. Abdominal trauma, lymphatic obstruction, inflammatory process, surgery, or radiation therapy are some causes of a secondary mesenteric lymphangioma.[3,4] The patients may be asymptomatic, or present with abdominal pain, due to compression of adjacent small bowel. Uncommonly they may present with acute abdomen due to torsion of the intestine around it, obstruction, secondary infection, rupture or hemorrhage.[5]

Mesenteric lymphangiomas are commonly an isolated disease process. To the best of our efforts, we could find only one case report in the available English literature published by Miele et al. in 2007, who reported the presence of cystic lymphangioma (CL) and hyposplenism in a case of celiac disease. They concluded in their article that although an association between CL and celiac disease cannot be suggested, a differential diagnosis of CL may be considered for cystic mesenteric masses in patients with celiac disease.[6] Mesenteric lymph node cavitation (MLNC) is seen in celiac disease and tuberculosis (TB). TB represents multiple caseating lymph nodes seen in small bowel mesentery and may be associated with other features of abdominal TB, like bowel wall thickening and ascites. Both of these carry different prognoses and treatment options with them. MLNC in celiac disease occurs late in the natural history of the disease and may represent an advanced disease, poor responsiveness to gluten free diet and carry a poor prognosis.[7] Empirical ATT for six to nine months, without a clinically defined endpoint of treatment, is often prescribed in peripheral healthcare facilities of many developing countries where tuberculosis is endemic to such patients without an affirmative diagnosis. Such treatment is not particularly beneficial to the patients and it should not be a routine practice.

Preoperative diagnosis of mesenteric cystic lesions can be accomplished by imaging with USG showing multiseptated cystic lesions and MRI helping to accurately determine the distribution and characterization.[8]

Conclusion

Mesentric lymphangioma may be rarely associated with other pathologies. A detailed clinical workup is necessary in each case guided by patient's clinical profile to give him maximum symptom relief.

Lessons Learned

Our case reports a young adult with multiple mesenteric lymphangioma with celiac disease, anemia, and autoimmune hepatitis. Apart from a rare combination of events, this case highlights the role of a detailed and appropriate work up before starting a patient on empirical antitubercular treatment especially in endemic countries.

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