Jejunal lymphangiectasia: A Rare Case Report

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Abstract: *Intestinal lymphangiectasia, characterized by dilatation of intestinal lacteals, is rare. The major treatment for primary intestinal lymphangiectasia is dietary modification. Surgical management is to be considered in presence of obstructive features, to relieve symptoms and to clarify the underlying etiology, also when medical treatment has failed or not possible. This article reports a 35 year old woman with segmental jejunallymphangiectasia, who presented with obstructive features- fullness of abdomen associated with epigastralgia, nausea and vomiting with history of loose motion alternating with constipation . Her symptoms are aggravated by taking food persisted with medical treatment. Surgery was finally performed to relieve the symptoms and to exclude the existence of underlying etiologies, with satisfactory effect. In conclusion, jejunal lymphangiectasia can present clinically asfullness of abdomen associated with epigastralgia, nausea and vomiting . Surgical management should be considered in presence of obstructive features to relieve pain and other symptoms and to exclude underlying diseases in some patients.*

I.   Introduction

Primary intestinal lymphangiectasia (PIL) is a rare disorder characterized by dilated intestinal lacteals resulting in lymph leakage into the small bowel lumen and responsible for protein-losing enteropathy leading to lymphopenia, hypoalbuminemia and hypogammaglobulinemia.Intestinal lymphangiectasia is a condition with widely variable symptoms and signs. Patients may be asymptomatic or present as vague abdominal pain, chronic diarrhea, steatorrhea, edemaPIL is also known as Waldmann's disease, In 1961, Waldmann et al. described the first 18 cases of "idiopathic hypercatabolic hypoproteinemia". These patients had edema associated with hypoproteinemia, low serum albumin and gammaglobulinlevels , Microscope examination of the small intestine biopsies showed variable degrees of dilation of the lymph vessels in the mucosa and submucosa. The authors also proposed the term "intestinal lymphangiectasia".The prevalence of clinically overt PIL is unknown. However, PIL can be asymptomatic; it primarily affects children (generally diagnosed before 3 years of age) and young adults but may be diagnosed later in dults Very rare familial forms of Waldmann's disease have been reported.The symptoms includes bilateral lower limb edema with anasarca and pleural effusion, pericarditis or chylous ascites. Fatigue, abdominal pain, weight loss, inability to gain weight, moderate diarrhea or fat-soluble vitamin deficiencies due to malabsorption may also be present.Patients complain of persistent diarrhea, abdominal pain, malabsorption, peripheral edema and chylous effusion. Obstructive ileus of the small intestine may develop, requiring partial jejunectomy[3]. In this condition, a sudden blockade of lymphatic drainage occurs in the affected area, followed by the massive dilation of submucosal channels and possible obstructive ileus. Congenital lymphedema is also associated with a selective deficit of naive CD4+ T lymphocytes. PIL with very low CD4+ counts and immunoglobulin G levels is related to recurrent and opportunistic infections and associated with increased morbidity and mortality.

Etiology remains unknown, The diagnosis of PIL is based on histological analysis of surgical specimens or endoscopic biopsies that reveal lacteal juice and dilated mucosal and submucosal lymphatic vessels. Typical mucosal findings upon endoscopy include diffuse swelling and enlarged whitish villi. Esophagogastroduodenoscopy and colonoscopy can be used to visualize parts of the small bowel, duodenum and terminal ileum, but capsule endoscopy is more useful to explore the entire small bowelmucosa. Double-balloon enteroscopy can localize lesions and small bowel tissue can be obtained for pathological confirmation. Differential diagnosis includes constrictive pericarditis, intestinal lymphoma, Whipple's disease, Crohn's disease, intestinal tuberculosis, sarcoidosis or systemic sclerosis. Several B-cell lymphomas confined to the gastrointestinal tract (stomach, jejunum, midgut, ileum) or with extra-intestinal localizations were reported in PIL patients.

A low-fat diet associated with medium-chain triglyceride supplementation is the cornerstone of PIL medical management. The absence of fat in the diet prevents chyle engorgement of the intestinal lymphatic vessels thereby preventing their rupture with its ensuing lymph loss. Medium-chain triglycerides are absorbed

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directly into the portal venous circulation and avoid lacteal overloading. Other inconsistently effective treatments have been proposed for PIL patients, such as antiplasmin, octreotide or corticosteroids.

Surgical small-bowel resection is useful in the rare cases with segmental and localized intestinal lymphangiectasia. The need for dietary control appears to be permanent, because clinical and biochemical findings reappear after low-fat diet withdrawal. PIL outcome may be severe even life-threatening when malignant complications or serous effusions occur.

Occasionally, IL can be seen in the aged people, which may be secondary to disorders causing lymphatic obstruction, such as lymphoma, carcinoma, tuberculosis, constrictive pericarditis, retroperitoneal fibrosis, post-radiation effects, and repeated parasite infestation. Diagnosis depends on characteristic endoscopic findings and pathological features[7-9]. However, it is sometimes difficult to differentiate primary from secondary IL. Surgical intervention may be a final resort to make a definite diagnosis and to relieve symptoms. Herein, we present a case with a segmental jejunal lymphangiectasia presenting asfullness of abdomen associated with epigastralgia, nausea and vomiting with history of loose motion alternating with constipation. The patient finally received surgical intervention and the symptoms resolved..

II. Case Report

A 35 year old woman from Turkmenistan was admitted to our hospital withobstructive features-fullness of abdomen associated with epigastralgia, nausea and vomiting with history of loose motion alternating with constipation. She had past surgical history of diagnosticlaparoscopy for the same complaints 2 months back in Iran.Investigations revealed lymphopenia, hypoalbuminemia, hyporglobulinemia, hypocalcemia and hypophosphatemia, cytopathology of peritoneal fluid shows lymphocytes, USG suggestive of thickened jejunum, CECT abdomen showed mural thickening of a segment of jejunum, capsule endoscopy was done which was suggestive of intestinal lyphangectasia of the segment of jejunum, and biopsy was taken which also showed lymphangiectasia of jejunum.
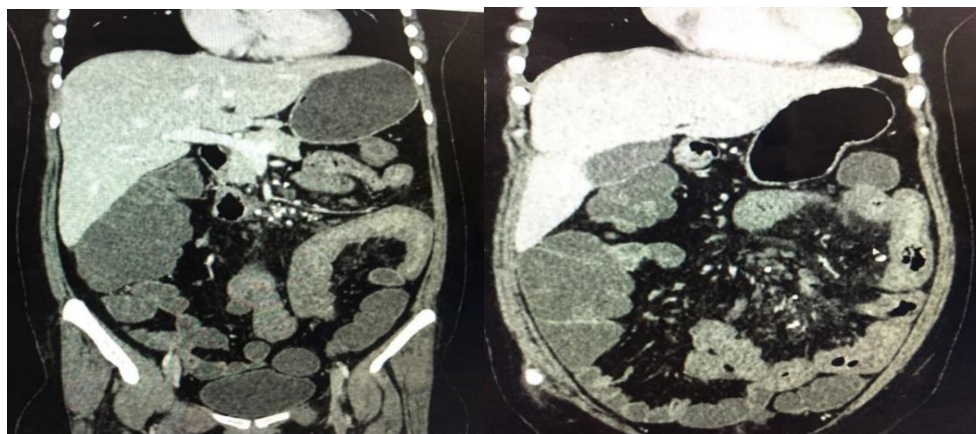
2.1 Ct Scan Enterography Showing-
1.   Long segment mural thickening involving the jejunum with inflamed jejunalmesentry and dialated stomach.
2.   Multiple prominent mesenteric lymph nodes with calcific foci.

Patient was taken up for surgical management and the final diagnosis of lymphangiectasia of jejunum is made by histopathological report of resected specimen of jejunum.

2.2Surgery
Patient was taken up for surgical management under general anesthesia,Midline laparotomy was performed on elective admission after written informed consent was obtained. After midline incision the peritoneal cavity was inspected the involved segment of the jejunum was identified clamped and resected then end to end anastomosis was performed along with paracolic drain placement.

Intra – operation findings
1.   300 ml of lymph was present in peritoneal cavity.
2.   Jejunal loops were distended with mural thickening.
3.   Multiple enlarged lymph nodes were present in mesentery
4.   Mesentery was thickened with dilated lymphatics.
5.   Intra-operative endoscopy was done to confirm the resection limit.

6. Around 5 feet of jejunum was resected 15cm distal to duodenojejunal flexure.
7. 8 feet of distal bowel proximal to ileocolic junction was preserved.
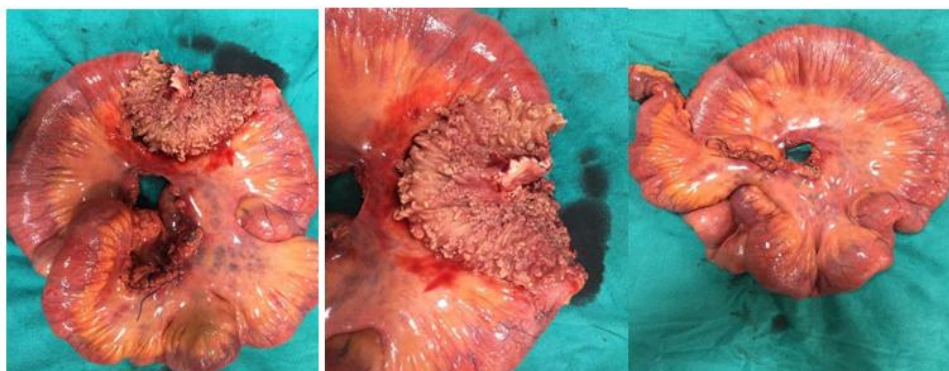
The resected segment was sent for histopathology and the laparotomy incision was closed in layers, the procedure time was 120 min, and no intraoperative complications occurred.

Post–operative period was uneventful and patientwas allowed orally on 5th post-operative day and drain was removed on 7th day. The patient was dischargedin stable condition on 8th post-operative day. Patient was completely asymptomatic, without any complications or recurrence and all the biochemical deraignment revert to normal.

III. Discussion

Intestinal lymphangiectasia has been well recognized as a disorder characterized by dilated lymphatic vessels of the gastrointestinal tract, especially the small intestine. It is a rare condition related to fat malabsorption and protein-losing enteropathy. Distribution of IL can be segmented, multifocal or diffuse. The pathogenesis is believed to be due to obstruction of lymphatic drainage. According to the etiologies, IL can be classified into primary and secondary forms. Primary IL is usually associated with many genetic syndromes, such as Turner's syndrome. On the contrary, secondary IL is acquired, due to several kinds of gastrointestinal diseases and intra-abdominal or retroperitoneal pathologies, such as carcinoma, lymphoma, tuberculosis or constrictive pericarditis.Clinical manifestations are similar in both forms of IL, but with variable severity according to the extent of involvement. Some patients can be completely asymptomatic, while at the other extreme, some may be associated with protein-losing enteropathy, growth retardation, or recurrent gastrointestinal tract bleeding. The protein loss is suspected to be due to rupture of the dilated intramucosal or submucosal lacteals, or exudation from the epithelium. The hemorrhage may be due to rupture of the dilated lacteals, which have potential communications with blood vessels.Diagnosis depends on clinical suspicion. Specific endoscopic findings, accompanied by typical histological pictures can draw into the diagnosis. These endoscopic findings include white-tipped villi, scattered white spots, white nodules, and submucosal elevations. Typical histological pictures consist of dilated intramucosal and submucosal lacteals. CT scan can help to find the underlying causes of secondary IL.Treatment of IL depends on the severity and extent of involvement. For most patients with primary IL, due to generalized abnormalities and diffuse distribution, dietary modification with a low-fat, high-protein diet and supplementation of medium-chain triglycerides (MCT) is the mainstay of treatment. As MCT is absorbed from the portal venous system directly rather than via lymphatics, it may avoid engorgement of the lymphatics, and thus reduce the opportunity for rupture. On the other hand, for patients with secondary IL, the underlying diseases should be treated. Surgical resection can be chosen when IL is confined to a segment of the intestine and has successfully treated protein-losing enteropathy, anemia or abdominal pain in intestine lymphangiectasia.

In this case, segmental involvement of jejunum withobstructive features-fullness of abdomen epigastralgia and nausea and vomiting with history of loose motion alternating with constipation was found. Biopsies showed a picture of IL, Due to segmental involvement and presence of obstructive features, also to rule out underlying causes, surgical resection was performed. The symptoms were relieved and a definite diagnosis of idiopathic jejunal IL was made.

IV. Conclusion

In conclusion, ThoughFor most the patients with primaryintestinal lymphangiectasia dietary modification with a low-fat, high-protein diet and supplementation of medium-chain triglycerides (MCT) is the mainstay of treatment but surgical management should be considered in presence of obstructive features to

relieve pain and other symptoms and to correct biochemical derangement, it also helps in excluding underlying diseases in patients with segmental jejunal lymphangiectasia.

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