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Case Report

Late onset Intestinal Lymphangiectasia -A rare cause of bipedal edema in middle age

Santokh Singh[*1], Gian Chand[2], Amit Verma[3] and Parampreet Singh[4]

[1]*Professor & Head, Department of Medicine & Gastroenterology, Govt. Medical College, Amritsar, India*
[2]*Associate Professor,Department of Medicine & Gastroenterology,Govt. Medical College, Amritsar, India*
[3]*Junior Resident, Department of Medicine & Gastroenterology, Govt. Medical College, Amritsar, India*
[4]*Intern, Department of Medicine & Gastroenterology, Govt. Medical College, Amritsar, India*

***Correspondence Info:**
Dr. Santokh Singh
9-FF HIG Flats A-Block
Ranjit Avenue Amritsar, India-143001
E-mail:: profsantokhsingh1@gmail.com

Abstract

Intestinal Lymphgiectasia is a rare disease characterized by hypoproteinemia, pedal edema and lymphocytopenia resulting from loss of lymph fluid into gastrointestinal tract. It can be primary or secondary and present during early childhood and rarely during adulthood. Presentation at 50 years is very very rare. The significance of this case lies in the late onset of Primary Intestinal lymphangiectasia which is a rare presentation of this very rare disease. The complaint of our patient was only bilateral pedal edema.

Keywords: Hypoproteinemia, Lymphocytopenia, Pedal edema, Dilated lacteals

1. Introduction

Bipedal edema is a common clinical presentation in outpatient department and routine causes include congestive heart failure, cirrhosis of liver, anemia with hypoproteinemia, chronic renal failure for pitting type of pedal edema and lymphatic obstruction & myxedema for the non pitting type of edema[1]. Sometimes rare diseases can present with common complaints making the diagnosis of the underlying disease difficult, as in this very case, which took long seven years to reach at final diagnosis though he visited many hospitals including secondary and tertiary level institution,which came only after intestinal biopsy in our institution.

2. Case report

A 50 year old male, nonhypertensive, nondiabetic, nonalcoholic presented in the medical OPD with complaint of progressively increasing swelling of both lower limbs for past seven years, which initially involved left lower limb and later right side. He denied any history of dyspnea, palpitation, chest pain, abdominal pain, cough with expectoration, jaundice, chronic weight loss, facial puffiness, cold intolerance, leg pains, chronic diarrhea, constipation and easy fatigability. There was no past history of jaundice, diarrhea, tuberculosis, renal ailment, heart failure, myxedema or visit to high endemic filariasis areas. He gave history of abdominal distension for the same duration and abdominal paracentesis being done once and said that the aspirate was milky white in color.

Examination revealed moderately built & nourished male, without evidence of anemia and signs of vitamin deficiency. Pulse rate was 80 per minute, regular, blood pressure 130/80 mmHg and JVP was not elevated. Bilateral pitting pedal edema was present more on the right side than the left (Figure 1). No signs of chronic liver disease or chronic renal

failure were present. Systemic examination did not reveal any lymphadenopathy, organomegaly or a lump in abdomen, no shifting dullness in abdomen. Lungs were clear and cardiac auscultation was essentially normal.

The previous investigations which the patient had with him of the last seven years were reviewed and showed TLC 8,400/cmm P78% L18 % M2% E2%, total serum protein 4.5 g/dl with serum albumin 2.4 g/dl and globulins 2.1 g/dl. Ascitic fluid analysis reported to be chylous in origin with protein content 1.3 g/dl.

Investigations done during the present admission showed Hb 11.5g/dl, TLC 10,600/cmm P83 % L15% E2%.Total serum protein 4.37 g/dl with serum albumin 1.6 g/dl serum globulin 2.77 g/dl. Liver and renal function test were normal. 24 hours protein in the urine was 18 mg/day. Ascitic tap revealed milky white ascitic fluid (chylous) exudative in nature with protein content 3.5 g/dl. USG abdomen showed normal sized liver and spleen with mild ascites. There was no other intra abdominal mass or lymph node enlargement. However the left kidney was hypoplastic and right kidney was normal. Doppler study of leg vessels revealed normal caliber and flow pattern in femoral veins, popliteal veins, anterior and posterior tibial veins and great and short saphenous veins. Saphenofemoral and saphenopopliteal were normal and no flow reversal seen on valsalva maneuver. Transthoracic echocardiography was essentially normal. Lymphangiscintigraphy had been done in tertiary level hospital by injecting 1 mCi Tc 99m sulphur colloid tracer into the web space of both feet revealed unobstructed lymphatic drainage in both limbs. Abdominal CT scan did not reveal any abnormality.

With this investigational workout, most of the common causes for pedal edema were ruled out. The significant finding of hypoproteinemia, in normal healthy non anemic person, on good nutritious diet and no loss of protein from the kidneys and normal liver functions made us think about possible loss of proteins from the gut. Further review of the literature showed that the presence of pedal edema, chylous ascitis, hypoproteinemia and lymphocytopenia may be a manifestation of intestinal lymphangiectasia. So an UGI endoscopy was performed which revealed normal looking mucosa in the esophagus and stomach. The duodenal & jejunal mucosa was edematous and a few white spots as evidence of dilated lymphatics were seen, biopsies were taken for HPE which revealed dilated sub mucosal lacteals consistent with diagnosis of intestinal lymphangiectasia. (Figure 2)

Figure 1-Left leg showing marked pedal edema

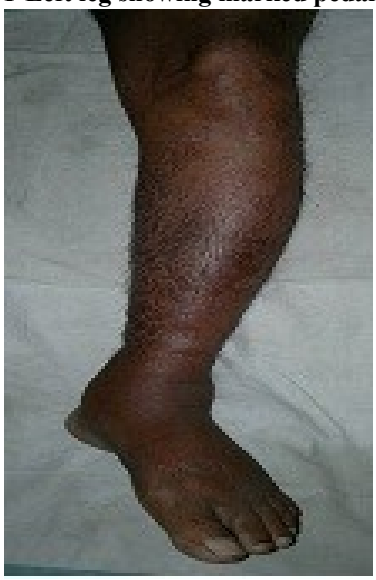
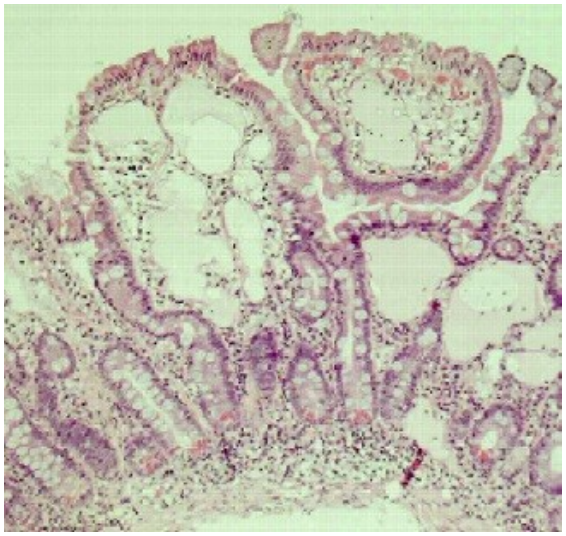


Figure 2 – Histopathology showing dilated lymphatics in the lamina propria of small bowel.



3. Discussion

Intestinal Lymphgiectasia is a rare disease characterized by hypoproteinemia, pedal odema and lymphocytopenia resulting from dilatation of intestinal lymphatics and loss of lymph fluid into gastrointestinal tract. It can be primary i.e. congenital or secondary due to other diseases e.g. secondary to constrictive pericarditis, intestinal lymphoma, lymphenteric fistula, Whipple's disease, Crohn's disease, sarcoidosis, intestinal tuberculosis, systemic sclerosis, radiation and/or chemotherapy with retroperitoneal fibrosis, human immunodeficiency virus-related enteropathy or the Fontan operation to treat cardiac malformations. Dilated lymphatics are located primarily in the small bowel resulting in severe leakage of lymph in to the gastrointestinal tract which translates into loss of proteins, immunoglobulins and lymphocytes[2,3].This entity

first described by Waldmann in 1961, primarily affects children (generally diagnosed before 3 years of age) and young adults but may be diagnosed later in adulthood[4,5]. Since then, no more than two hundred cases have been reported[6]. Presentation in children is with non bloody diarrhea and odema. Malabsorption, steatorrhea, lymphocytopenia and hypoalbuminemia can be other presenting complaints. Chylous ascites and chylous pleural effusion are also reported in patients with long standing lymphangiectasia[2,7]. Various vitamin deficiencies may be seen and osteomalacia from vitamin D deficiency was also reported[7].

Endoscopic findings in PIL are edema of duodenal mucosa and creamy yellow coloured jejunal villi corresponding to marked dilatation of the lymphatics within intestinal mucosa. Endoscopy may be negative when intestinal lesions are localized or segmented. In such cases, videoscope enteroscopy may be used. Histopathological examination of duodenum-jejunum and ileum biopsies confirm the presence of lacteal juice dilated mucosa (from moderate to severe) and submucosal lymphatic vessels with polyclonal normal plasma cells[8].CT and MRI are helpful in rulling out many secondary forms of intestinal lymphangiectasia. CT may show dilatation of small bowel, thickening and hypervascularity of the mucosal folds with MRI confirming the hyperintensity on T1 images suggestive of protein rich fluid[9,10].

Various modalities have been used to treat this condition, but with limited success – low fat diet with supplementary medium chain triglycerides, antiplasmin – tranexamic acid, octreotide, surgical resection and steroids[3,6].

In our patient, the presence of bipedal edema with lymphopenia and chylous ascites with other common causes ruled out by clinical history, examination and investigations suggests a diagnosis of intestinal lymphangiectasia . CT scan in our patient was normal and scintigraphy was normal at a tertiary care centre (perhaps due to intermittent nature of protein loss)[11]. Endoscopic biopsy of duodenum, jejunum showed presence of dilated lacteals which confirmed intestinal lymphangiectasia. Taking into consideration all the investigations in this case, secondary causes of intestinal lymphangiectasia are effectively ruled out giving the final impression of late onset primary intestinal lymphangiectasia.

4. Conclusion

Intestinal lymphangiectasis mostly present during childhood and less commonly during adulthood, presentation at 50 years is very very rare. Most of the literatures reviewed have described case during childhood and adults. The significance of this case lies in the late onset of Primary Intestinal lymphangiectasia which is a rare presentation of this very rare disease.

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