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**Case Report**

# Intestinal lymphangiectasia: case report

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## ABSTRACT

Primary intestinal lymphangiectasia (PIL) was first described by Waldmann et al, in 1961. PIL is a rare disease with several hundred reported cases. It is rarely reported in adults because it is presumably a congenital disorder and when present in adults it typically produces a long duration of manifestation such as diarrhea, abdominal distention from ascites, and peripheral edema. This disorder is characterized by markedly dilated intestinal lymphatics, hypoproteinemia, generalized edema, lymphocytopenia hypogammaglobinemia, and immunologic anomalies. The loss of protein into the from dilated intestinal lymphatics leads to the development of hypoproteinemia in these patients and its demonstration is important in the diagnosis of intestinal lymphangiectasia. The disease can be secondary to congenital, secondary or idiopathic defects in the formation of the lymphatic ducts. In the present report, we describe a case of 15 years old female presented to our hospital with history of generalized edema, bilateral hand spasm, and diarrhea. Endoscopy of the patient revealed White spots (dilated lacteals), white nodules, and submucosal elevations were observed. Changes suggestive of the disease includes White villi and/or spots (dilated lacteals), white nodules, and submucosal elevations are observed. Xanthomata's plaques are often visualized, there are no specific treatments for patients with PIL. treatment of patients with primary intestinal lymphangiectasia involves control of symptoms with the use of dietary, pharmaceutical, and behavioral modifications.

**Keywords:** Dietary, Intestinal, Lymphangiectasia, Rare, Villi

## INTRODUCTION

Primary intestinal lymphangiectasia (PIL) was first described 1961.[1] PIL is a rare disease with several hundred reported cases.[2] It is rarely reported in adults because it is presumably a congenital disorder and when present in adults it typically produces a long duration of manifestation such as diarrhea, abdominal distention from ascites, and peripheral edema.[3] This disorder is characterized by markedly dilated intestinal lymphatics, hypoproteinemia, generalized edema, lymphocytopenia hypogammaglobinemia, and immunologic anomalies.[4] The loss of protein into the from dilated intestinal lymphatics leads to the development of hypoproteinemia in these patients and its demonstration is important in the diagnosis of intestinal lymphangiectasia.[5]

Authors experienced a case of primary intestinal lymphangiectasia that was confirmed by endoscopic intestinal biopsy showing markedly dilated intestinal lymphatics.

## CASE REPORT

### *Patient information*

15 years old girl referred to our hospital from secondary hospital as case of protein losing enteropathy for further evaluation and management.

Presented to our ER with history of generalized edema, bilateral hand spasm, and diarrhea for one week.

She was in her usual state of health till 3 days back when she started to have morning bilateral periorbital edema that decreases at the end of the day with bilateral lower limb edema reaching above her knees, and abdominal distension which was progressive in nature with no specific aggravating or relieving factors.

The swelling was not associated with pain, skin changes and disabling her form daily activity.

Along with swelling she had bilateral hand spasm which started suddenly and progressed over time, painful to the degree that she was not able to close her hands.

Also, she complained from perioral numbness and her mother noticed mild facial twitching started one hour before the hand spasm (eye blinking and checks twitching).

On the 3rd day she developed diarrhea, which was watery to semi-solid stool, 7 times/day, not bloody with no mucus noted. There was history of mild on and off abdominal pain that is relieved by defecation.

The patient was following in in private hospital for 6 months when her symptoms started in which she was labeled as case of protein losing enteropathy with unknown etiology after non conclusive endoscopy. Having multiple episodes of similar presentation every 2 to 3 weeks.

Requiring admission for albumin transfusion and electrolyte imbalance.

***Clinical findings***

Examination revealed Weight: 42.50 KG on 49th centile. Height: 157CM on 75th centile, No dysmorphic features. The patient had positive Chvostek's sign. Bilateral lower limb pitting edema reaching to above her knees. No nails/ skin change, pulses are intact. Other examinations were unremarkable.

***Diagnostic assessment***

The labs of the patient showed lymphocytopenia, hypocalcemia, hypomagnesemia, hypoalbuminemia and hypogammaglobulinemia (Table 1).

The Differentials diagnosis that considered were constrictive pericarditis, heart failure, retroperitoneal fibrosis, abdominal tuberculosis, retroperitoneal malignancy, Nephrotic syndrome and Celiac disease.

Endoscopy of the patient revealed White spots (dilated lacteals), white nodules, and submucosal elevations were observed (Figure 1) and biopsy were taking.

**Table 1: Lab values of the patient.**

| Parameter | Value | Normal |
|---|---|---|
| WBC | 4.00 | 4.30-11.30 |
| RBC | 3.33 | 4.30-5.50 |
| Lymphocyte A | 0.51 | 1.90-4.90 |
| Ca | 0.99 | 2.10-2.60 |
| Corrected calcium | 1,4 | |
| Mg | 0.33 | 0.70-1 |
| Albumin | 14 | 40-50 |
| Total protein | 354 | 65-81 |
| IgG | <3.0 | 7-16 |
| IgA | <0.50 | 0.70-4 |
| IgM | <0.31 | 0.40-2.30 |
| Globulin | 14 | 20-39 |

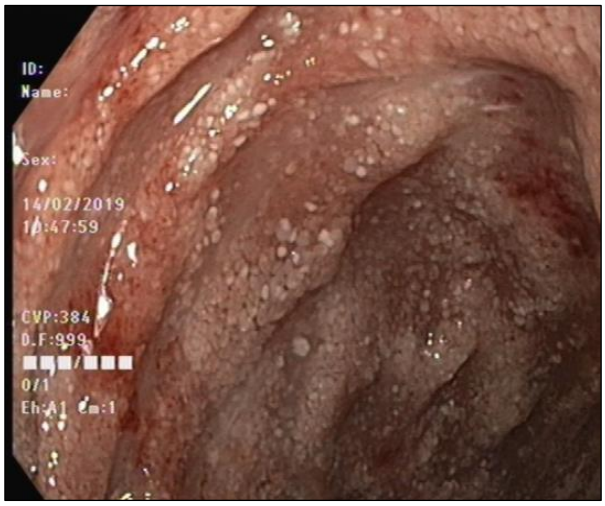


**Figure 1: Endoscopy image showing white spots (dilated lacteals).**

The specimen was sent for histopathological examination which revealed Dilation of mucosal and submucosal lymphatic channels.

***Therapeutic intervention***

Authors elect to use medical treatment as the patient is responsive to it , patient received albumin replacement and dietary modification in addition to the use of octreotide. patient did well during admission and discharged with close follow ups every 2 months for the first year then biannual visits.

## DISCUSSION

Primary intestinal lymphangiectasia (PIL) was first described in 1961.[1] PIL is a rare disease with several hundred reported cases.[2] It is rarely reported in adults because it is presumably a congenital disorder and when present in adults it typically produces a long duration of manifestation such as diarrhea, abdominal distention from ascites, and peripheral edema.[3] This disorder is

characterized by markedly dilated intestinal lymphatics, hypoproteinemia, generalized edema, lymphocytopenia hypogammaglobinemia, and immunologic anomalies.[4] The loss of protein into the from dilated intestinal lymphatics leads to the development of hypoproteinemia in these patients and its demonstration is important in the diagnosis of intestinal lymphangiectasia.[5]

The disease can be secondary to congenital, secondary or idiopathic defects in the formation of the lymphatic ducts. Congenital forms are frequently associated with Turner's, Noonan's and Klipper-Trenaunay-Weber Syndromes. Secondary forms are associated with constrictive pericarditis, heart failure, retroperitoneal fibrosis, abdominal tuberculosis, retroperitoneal malignancy and other pathologies.[6]

Various methods have been used to diagnose this disorder, including α1-antitrypsin intestinal clearance test and tests using radiolabeled macromolecules, have been used to detect gastrointestinal protein losses. Imaging Studies such as double-contrast radiographs of the small bowel may be helpful, as it may show thickened folds due to intestinal edema from protein losing. Furthermore, ultrasonography and computed tomography (CT) scanning are also useful in identifying dilated intestinal loops.[7,8] Repeatedly, the role of endoscopy has been proven useful.[9] Small bowel enteroscopy not only helps detect mucosal changes suggestive of the disease but also allows acquisition of histologic samples to establish a diagnosis.[9,10]

changes suggestive of the disease includes White villi and/or spots (dilated lacteals), white nodules, and submucosal elevations are observed. Xanthomatous plaques are often visualized.[11]

There are no specific treatments for patients with PIL. treatment of patients with primary intestinal lymphangiectasia involves control of symptoms with the use of dietary, pharmaceutical, and behavioral modifications. Segmental resection may be recommended But, in over half of the cases, treatment with a high-protein, fat-free diet, and supplementation of medium-chain triglyceride that is absorbed directly into the portal system without development of lacteal engorgement, is usually effective in preventing and alleviating the symptoms.[12,13]

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