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CASE REPORT

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A late diagnosis of primary intestinal lymphangiectasia in a Syrian girl

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Key Clinical Message

Primary intestinal lymphangiectasia is a rare disease that affects children and young adults, causing mainly gastrointestinal disorders that lead to edema and immunologic abnormalities. The majority of patients typically present bilateral lower limb edema, along with pericarditis, pleural effusion, or chylous ascites. The goal of the treatment is to restrict the consumption of long-chain fats and administer a formula that includes protein and medium-chain triglycerides. Here, we report 11-year-old Syrian girl presented to the hospital with 2 years history of diarrhea and generalized edema. Furthermore, investigation showed ascites and pleural effusion. The disease was detected by several biopsies from duodenum that showed dilation of the lymph vessels within the lamina propria without any evidence for inflammation. She was managed by modifying her diet and albumin transfusion, which caused significant improvement in the child's condition.

KEYWORDS

case report, edema, gastrointestinal, primary intestinal lymphangiectasia

1 | INTRODUCTION

Intestinal lymphangiectasia is an uncommon non-cancerous disease characterized by the expansion of the lymphatic vessels in the mucosal, submucosal, and subserosal layers of the intestines. It is considered a significant factor contributing to protein loss in the digestive system.[1]

There are two types of intestinal lymphangiectasia, namely primary and secondary. Therefore, the diagnosis of primary intestinal lymphangiectasia (PIL) must first exclude the possibility of secondary intestinal lymphangiectasia.[2] The primary symptom of this condition is primarily bilateral lower limb edema, which can vary in severity from moderate to severe, and may also involve anasarca, and it could be accompanied by other complications such as pleural effusion, pericarditis, or chylous ascites.[2]

PIL is generally diagnosed before the age of three but it may be diagnosed in older patients.[3] There is no exact information about the incidence and prevalence of this disorder.[4] About 200 cases of PIL were reported in the literature, and the vast majority of patients were diagnosed before the third year of life.[5]

Here, we reported a successful treatment of 11-year-old Syrian girl who presented with 9 years history of diarrhea and generalized edema.

2 | CASE PRESENTATION

An 11-year-old Syrian girl had been admitted to Aleppo University Hospital with a history of eyelids swelling, generalized edema, and abdominal distension. The patient has had a history of intermittent diarrhea since she was 2 years old and history of oligouria for the past 2 months in addition to fatigue and poor oral intake. She arrived late to hospital due to the conditions of war.

She had adhered to a gluten-free diet as recommended by pediatricians without clinical response. The physical examination showed normal vital signs; the weight and height were normal for her age.

The abdomen was distended with evidence of shifting dullness, and we also noticed anasarca and few hyperpigmented patches over the face and neck. By auscultation of chest: ↓A/E equally at the bases of both lungs, the examination of the other systems was normal.

Laboratory tests showed lymphocytopenia (lymphocytes 8.7%), hypoproteinemia (total protein 27.5 g/L), hypoalbuminemia (serum albumin 14 g/L), hypoimmunoglobulinemia (serum globulin 1.35 g/L), and hypolipoproteinemia (cholesterol 2356 mmol/L, TG: 0.6667 mmol/L). The other laboratory values were normal.

By paracentesis abdominis, the fluid was chylous. The patient underwent several radiology investigations (US, MS-CT, Endoscopy).

US abdomen demonstrated massive ascites and pleural effusion, and echocardiography showed mild pericardial effusion. MS-CT (chest, abdomen and pelvic) revealed bilateral pleural effusion, pericardial effusion, atelectasis, and massive fluid in abdomen and pelvic.

Upper gastrointestinal endoscopy was normal even though several biopsies were taken from duodenum, one of which showed intestinal lymphangiectasia with non-specific duodenitis (Figure 1).

The patient was treated with albumin transfusion, low-fat regimen, high protein, and fat-soluble vitamins. She responded to this treatment and had remarkable improvement in her symptoms, where diarrhea and edema gradually subsided and she started to gain weight.

After 5 years of follow-up, the patient showed a good response to the treatment with no recurrence.

3 | DISCUSSION

Intestinal lymphangiectasia is a relatively rare disorder that manifests itself through-intestine disorder malabsorption.[2] It may affect both males and females equally.[1]

Intestinal lymphangiectasia can be categorized into primary or secondary forms, which can be caused by various underlying conditions such as sarcoidosis, lymphoma, constrictive pericarditis, and scleroderma.[1]

Primary intestinal lymphangiectasia is caused by morphologic abnormalities (genetic mutations)[2] which lead to obstruction and dilation of lymphatic vessels in the intestines. So, the patient loses proteins especially albumin and gammaglobulin, lymphocytes, and other

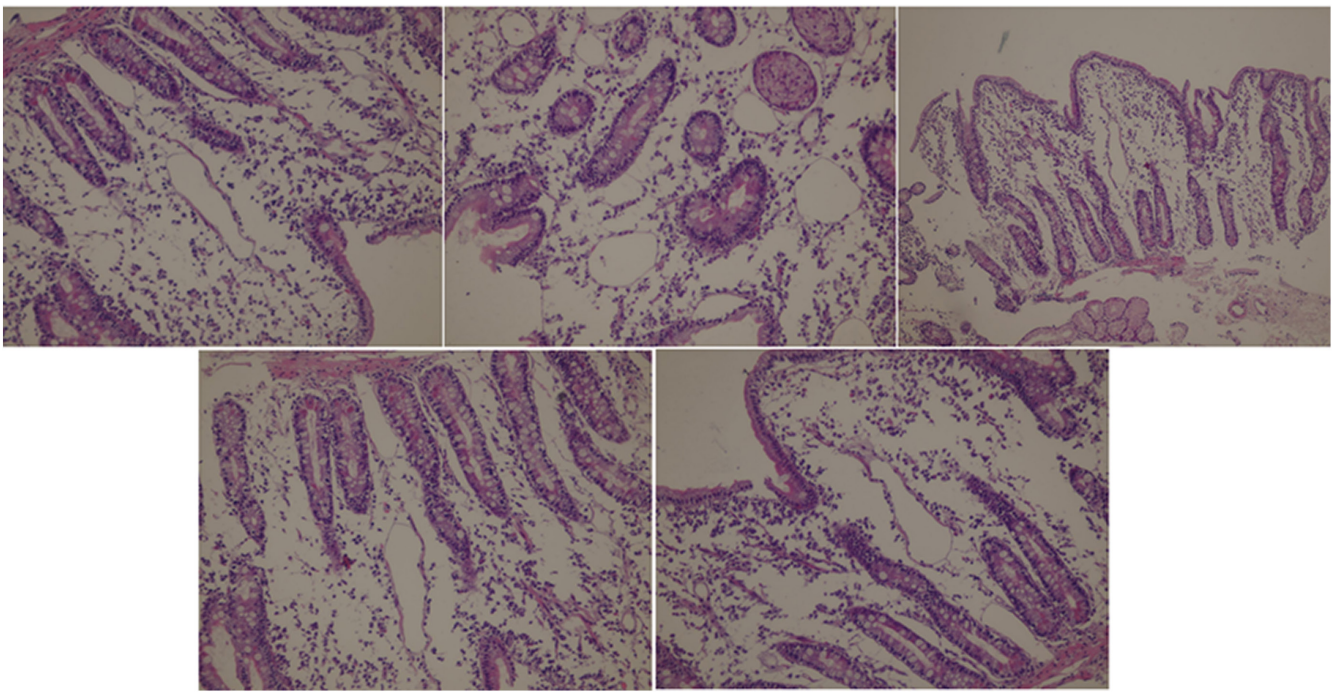

FIGURE 1 Intestinal lymphangiectasia with non-specific duodenitis.

serum components (lipid and fat-soluble vitamins) in the lumen of the intestine.[1]

Clinical manifestations of PIL are intermittent peripheral edemas, steatorrhea, abdominal pain, and ascites.[4]

Radiological tools could help in diagnosis; we can see pleural effusion and ascites. Upper gastrointestinal endoscopy usually shows whitish patchy distribution or creamy yellow of jejunal villi corresponding to marked dilation of lymphatics,[3,6] but in our case it was normal, so the diagnosis was confirmed by small bowel biopsy. With our patient, biopsies showed duodenal mucosa with a moderate degree of villous shortening and broading associated with increased intraepithelial lymphocytes and plasma cells, normal muscularis mucosa, brunner glands, and presence of dilated lymphatic channels within the lamina propria.

The goal of treatment is to limit the intake of long-chain fats and administering a formula that includes protein and medium-chain triglycerides.[6] In cases where a low-fat and medium-chain triglycerides diet is not effective, medical therapy such as antiplasmin therapy and the use of octreotide, a medication similar to somatostatin, may be recommended.[1]

4 | CONCLUSION

PIL is a rare disease that typically develops in early childhood. As the symptoms of PIL resemble those of other diseases, it can be misdiagnosed or diagnosed late, as in the case we presented.

Therefore, we recommend considering PIL in children who present with edema or diarrhea with lymphocytopenia, even if not all features are obvious. A small intestine biopsy can help confirm the diagnosis of PIL.

The most effective management of PIL is the application of a low-fat diet and substituting long-chain fatty acids with medium-chain fatty acids. Further studies are needed to determine the prevalence and incidence of PIL, and to develop more effective management strategies for this rare disease.

AUTHOR CONTRIBUTIONS

Afaf Mohammad Maher Najjar: Conceptualization; data curation; investigation; writing – original draft; writing – review and editing; guarantor. **Nour khaled babensi:** Conceptualization; data curation; formal analysis; project administration; validation; writing – original draft. **Ahmad ghazal:** Investigation; supervision; validation; writing – original draft. **Muhamad Zakaria Brimo Alsaman:** Writing – original draft; writing – review and editing. **Alaa Aldin Ismail**: Supervision; writing – review and editing. **Hasnaa Alnaeb:** Project administration; supervision; writing – original draft.

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There were no sources of funding for this case.

CONFLICT OF INTEREST STATEMENT

None declared.

DATA AVAILABILITY STATEMENT

Data sharing is not applicable to this article as no datasets were generated or analyzed during the current study.

ETHICAL APPROVAL

No ethical approval was required for this case report.

CONSENT

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

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