

Exacerbation of primary intestinal lymphangiectasia during late pregnancy and recovery after delivery

A case report and literature review

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Abstract

Rationale: Primary intestinal lymphangiectasia (PIL) is a rare disease characterized by dilated intestinal lacteals resulting in lymph leakage into the small bowel lumen. Main clinical features include intermittent diarrhea, hypoproteinemia. Scattered case reports suggested that PIL is compatible to pregnancy, but with increased complications.

Patient concerns: A 34-year-old woman with endoscopically diagnosed PIL presented to antenatal our clinic at 10 weeks into gestation. She reported strict adherence to low-fat/high-protein diet with medium-chain triglycerides (MCTs) supplementation. She was general well except for moderate edema and hypoalbuminemia. At 33 weeks, she developed diarrhea, nausea, and vomiting, with decreased fetal movements. One week later, she had an asthma attack. Nonstress test showed frequent variable deceleration.

Diagnoses: The diagnosis of PIL was established endoscopically 8 years earlier.

Interventions: Hypoalbuminemia was corrected with intravenous albumin administration. She also received corticosteroid therapy to promote fetal lung maturation in anticipation to early termination of the pregnancy.

Outcomes: A cesarean section was carried out at 34 weeks due to fetal distress. The baby girl was apparently healthy: weighing 2160g, with an Apgar score of 9 at both 1 and 5 minutes. Symptoms dissipated rapidly after the delivery. The last follow-up visit at 15 months was unremarkable for both the mother and infant.

Lessons: PIL could be compatible with pregnancy, but requires strict adherence to dietary treatment, proper management of the symptoms (e.g., hypoalbuminemia), particularly during late gestation.

Abbreviations: MCTs = medium-chain triglycerides, NST = nonstress testing, PIL = primary intestinal lymphangiectasia.

Keywords: hypoalbuminemia, pregnancy, primary intestinal lymphangiectasia

1. Introduction

Primary intestinal lymphangiectasia (PIL), first described by Waldmann in 1961,^[1] is a rare disease characterized by dilated intestinal lacteals resulting in lymph leakage into the small bowel lumen. Major clinical features include lymphopenia, hypoalbuminemia, and hypogammaglobulinemia.^[2-4] Other symptoms include nausea/vomiting, weight loss, chronic fatigue, edema, intermittent diarrhea, steatorrhea, and abdominal pain. Fat-soluble vitamin deficiencies may occur. Symptoms typically emerge in childhood; cases with adult onset have only been occasionally reported. Definitive diagnosis is established on the

basis of endoscopic examination of the small bowel. Mainstay treatment consists of long-term low-fat diet and supplementation of medium-chain triglycerides (MCTs).^[2,3] Here, we report a case of a PIL with childhood onset. Symptoms exacerbated during late pregnancy and dissipated after delivery.

2. Case report

A 34-year-old woman started attending antenatal clinic 10 weeks into pregnancy. Stating from infancy, she had intermittent diarrhea, edema, and occasional tetany. She also had asthma and is allergic to erythromycin and aminophylline. A diagnosis of PIL was established by endoscopy (Fig. 1) in the First Affiliated Hospital, Zhejiang University 8 years ago (at an age of 26). She was then placed on a low-fat/high-protein diet with MCTs supplementation. Compliance was reasonably good. She became pregnant 3 years earlier; antenatal clinical visit noticed subclinical hypothyroidism, but she did not receive replacement therapy, and the pregnancy ended with miscarriage. For the current episode of pregnancy, subclinical hypothyroidism was confirmed again (Table 1), and she received levothyroxine treatment (125 mg daily).

The antenatal clinical visit was conducted at a frequency of once every 2 weeks. She had persistent edema in both legs and eyelids. Serum albumin was between 20 and 24 g/L (normal reference range, 40–55 g/L). Urine examination did not show proteinuria.

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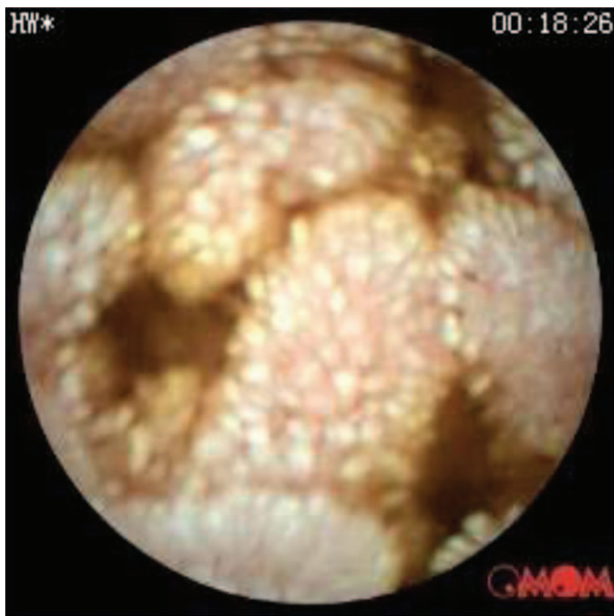


Figure 1. Capsule endoscopy showing lymphangiectasia in the small intestine.

33 weeks into the pregnancy, she developed diarrhea, nausea, and vomiting, and was admitted for decreased fetal movements. Blood pressure was normal (129/69 mmHg). A nonstress test showed fetal distress. A physical examination revealed pitting edema in both legs and eyelids. Weight gain during the pregnancy was 12.5 kg. Body mass index was 25 kg/m² (height: 160 cm; weight: 64 kg). Fundal height was 30 cm. Abdominal circumference was 95 cm. Fetal ultrasound showed biparietal diameter at 8.0 cm, and femur length at 6.1 cm.

Blood routine showed normal white blood cell count (8900/mm³), with a differential count of 96.5% neutrophils (normal range, 40–75%), and 1.6% lymphocytes (normal range, 20–50%). Hemoglobin was 13.8 g/dL with normal hematocrit (40.7%). Platelet count was 251,000/mm³. Serum total protein was 42.2 g/L, with hypoalbuminemia (albumin, 19.6 g/L) (Table 1). Urine routine did not reveal proteinuria. Stool occult blood was negative. Liver and renal function tests were normal.

Table 1

Blood routine examination and biochemistry panel.

Blood test	Results	Normal range
WBC and differential	8900 (N 96.5%–L 1.6%)	3.5–9.5 × 1000/mm ³
HGB	13.8	11.5–15.0 g/dL
HCT	40.7	35–45%
PLT	251	125–350 × 1000/mm ³
CRP	14	mg/L
Urea	6.36	3.1–8.8 mmol/L
Creatinine	49	41–111 μmol/L
ALT/AST	10/29	7–40 U/L
PT	10.1	seconds
INR	0.86	
Calcium	1.72	2.0–2.6 mmol/L
Magnesium	0.53	0.66–1.07 mmol/L
Total protein	42.2	65–85 g/L
Albumin	19.6	40–55 g/L
TSH	3.354	0.35–5.5 mIU/L
FT3	4.78	3.5–6.5 pmol/L
FT4	19.89	11.5–22.7 pmol/L
Cholesterol	5.03	2.85–5.7 mmol/L
LDL	2.58	1.31–3.37 mmol/L
HDL	1.15	0.9–1.95 mmol/L
TG	4.43	0.56–1.7 mmol/L

ALT = alanine aminotransferase, AST = aspartate aminotransferase, CBC = cell blood count, CRP = C-reactive protein, FT3 = free triiodothyronine, FT4 = free thyroxine, HCT = hematocrit, HDL = high-density lipoprotein, HGB = hemoglobin, INR = international normalized ratio, LDL = low-density lipoprotein, LFT = liver function test, PLT = platelet, PT = prothrombin time, RFT = renal function test, TG = triglycerides, TSH = thyroid stimulating hormone, WBC = white blood cell.

Ultrasound examination of the chest and abdominal cavities and organs was unremarkable.

The patient received dexamethasone (6 mg, i.m., every 12 hours for a total dose of 24 mg) to promote fetal lung maturation in anticipation to early termination of the pregnancy. She also received 5 10 mg doses of intravenous (IV) albumin every 24 hours to correct hypoalbuminemia (albumin, 19.6 g/L elevated to 25.3 g/L). One week after hospitalization (34 weeks of pregnancy), she had an asthma attack. Based on nonstress test (Fig. 2), cesarean section was performed. She delivered an apparently healthy girl, weighing 2160 g, with an Apgar score of 9 at both 1 and 5 minutes. Edema in the legs dissipated over a period of 3 days after the C-section. Hypoalbuminemia, however, persisted (albumin at 20.3 g/L and total proteins at 42.3 g/L upon discharge



Figure 2. Frequent variable deceleration at 34 weeks of gestation.

Table 2**Summary of the 7 reported cases of PIL with pregnancy.**

Ref.	Clinical features	Pregnancy outcome
Liu and Sherman, ^[5] Ghoshal et al, ^[6]	Subconjunctival edema, hypoglobulinemia. Lymphopenia, hypoalbuminemia.	Uneventful, a normal vaginal delivery occurred at 40 weeks of gestation. The patient delivered a full-term, low-birth-weight infant, who died a day after birth. Diagnosis of PIL was confirmed in postpartum period.
Voigt et al, ^[7] Quemere et al, ^[8]	Article in German. Hypoalbuminemia, edema.	A healthy, 3400 g female infant was born near term by cesarean section. The patient delivered a full-term infant by forceps and experienced no major complications.
Tourlakis et al, ^[9]	Hypoalbuminemia, edema.	The patient received intravenous albumin at regular intervals and gave birth by cesarean section due to placenta previa to a healthy female fetus at 39 weeks of gestation.
Szucs et al, ^[10] Ibarguengoitia et al, ^[11]		Article in Hungarian, no abstract. Article in Spanish, no abstract.

PIL = primary intestinal lymphangiectasia.

on 6 days after the C-section). Follow-up of the patient indicated that edema aggravated upon exclusive breast-feeding and alleviated significantly upon mixed feeding. The last follow-up at 15 months was not remarkable.

3. Discussion

PIL is a rare disease that typically manifests itself in childhood. The underlying pathophysiology and clinical features of this disease apparently overlaps with the physiological changes that happened during pregnancy (e.g., hormonal changes and the resulting fluid retention). However, only 7 cases of pregnancy of PIL patients have been reported. The characteristics of the 7 cases are summarized in Table 2.^[5–11] Based on these reports, we believe that PIL is generally compatible with pregnancy, but requires heightened alert. In the current case, the patient was generally well except edema and hypoalbuminemia until 33 weeks into the gestation, when diarrhea, nausea, and vomiting emerged. The timing is apparently consistent with the rapid blood volume increase in 32 to 34 weeks of gestation. Another factor could be the rapidly increasing metabolic needs and food intake that in turn place additional burden onto deformed lymphatic vessels. Symptoms of nausea and vomiting, together with hypoproteinemia, could jeopardize nutrition supply to the developing fetus. Ghoshal et al^[6] reported a low-birth-weight of an infant by a mother with PIL diagnosis established postpartum. In the current case, hypoproteinemia was managed with intravenous albumin. This may have contributed to the normal development of the fetus, as noted previously by Tourlakis et al.^[9] Other symptoms of PIL, including weight-loss, iron deficiency anemia, fat-soluble vitamin deficiency,^[12,13] could also complicate pregnancy if not managed properly. Dietary therapy is the cornerstone of PIL management.^[14] Based on self-report, the patient in the current case strictly adhered to the low-fat/high-protein diet with supplementation of MCTs. We believe that good compliance may have contributed to the successful pregnancy and outcome to both the mother and infant in this case.

4. Conclusion

PIL could be compatible with pregnancy, but requires strict adherence to dietary treatment, proper management of the

symptoms (e.g., hypoalbuminemia), particularly during late gestation.

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