Fatty gut needs low-fat formula

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To the Editor: 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.[1] Etiology and prevalence of the PIL are unknown. PIL affects males and females equally, and there is no racial predisposition. There is no specific treatment for PIL and fat restriction forms the cornerstone of treatment.[1] There is very little case reported in Asia.

In May 2016, a 5-month-old baby, who presented with chronic diarrhea and severe failure to thrive since the age of 1 month. The mother describes diarrhea as cheesy and foully smelling. Diarrhea (ten loose bowel movements per day) is associated with significant weight loss. Physical examination reveals that her weight and height are both below the fifth percentile for age. He was wasted and there were no subcutaneous fat tissues [Figure 1]. The laboratory study findings include a hemoglobin level of 102 g/L (reference range, 122–153 g/L) and serum albumin of 19 g/L (reference range, 35–55 g/L). White blood cell, platelet, C-reactive protein, bilirubin, alkaline phosphatase, alanine transaminase, and aspartate aminotransferase are normal. The stool sample contains many fat droplets, but his stool culture results are negative. Random stool for alpha-1-antitrypsin is 3.7 mg/g (reference range, 0–0.5 mg/g). Esophagogastroscopy reveals edematous duodenal mucosa with scattered whitish spots. The histopathologic biopsy of the duodenum reveals dilated lymphatics [Figure 2]. A diagnosis of PIL is made based on history, physical examination, hypoalbuminemia, positive stool alpha-1 antitrypsin, and corroboration on histopathology findings.

We started our patient on medium-chain triglycerides (MCT) based diet (Monogen^{R} formula) and fat restriction. Monogen is a nutritionally complete, low fat, whole whey protein powdered feed, low in long-chain triglycerides and high in MCT and supplemented with docosahexaenoic acid and arachidonic acid. The need for dietary control appears to be permanent, because clinical and biochemical findings reappear after low-fat diet withdrawal. After 3 months he gained weight, and diarrhea improved [Figure 3]. On office follow-up after hospital discharge, laboratory findings (serum albumin and stool alpha-1-antitrypsin) were normalized.

PIL is an extremely rare disorder characterized by dilated intestinal lacteals resulting in lymph leakage into the small bowel. Depending on the cause of the disease, it can be classified into primary PIL, which can occur as an isolated disorder (from birth) or as a part of syndromes such as Noonan, Klippel-Trennay-Weber, von Recklinghausen, Hennekam, and yellow nail syndrome.[2] Secondary intestinal lymphangiectasia occurs as a complication to disease states, such as constrictive pericarditis, lymphoma, sarcoidosis, scleroderma, Whipple's disease, and Crohn's disease.[3]

There is no specific treatment for PIL and fat restriction forms the cornerstone of treatment.[4] It is likely that the absence of fat in the diet prevents engorgement of the intestinal lymphatics with chyle, thereby preventing their rupture that results in protein and T-cell loss. Thus, a low-fat diet with supplemental MCT forms the cornerstone of management in PIL. MCT are directly absorbed into the portal venous system, which prevents lacteal engorgement. Small bowel resection may be helpful if the disease involves a localized segment of bowel.[5] Octreotide, antiplasmin (tranexamic acid), and corticosteroids are treatment options described in the literature, but there is insufficient data for their use.[5]

Declaration of patient consent

The patient was a participant of our registered clinical trial which was approved by Institutional Ethical of King Saud Bin Abdulaziz University for Health Sciences, and informed consent was achieved from the patient.

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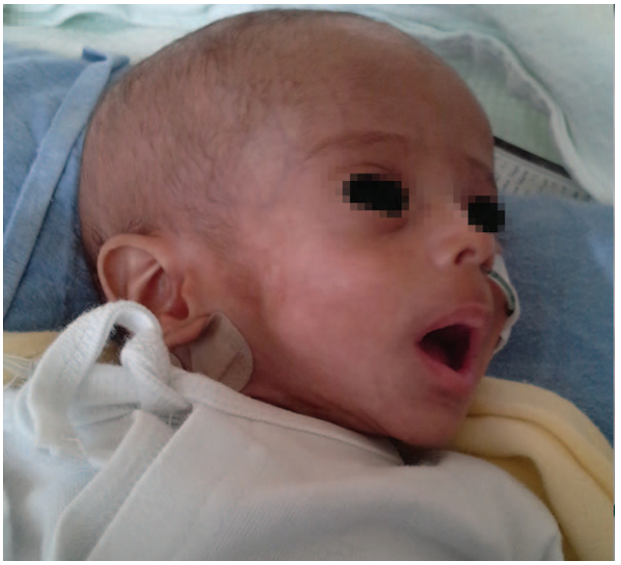

Figure 1: A wasted infant with loss of subcutaneous fat tissues (pre-treatment).

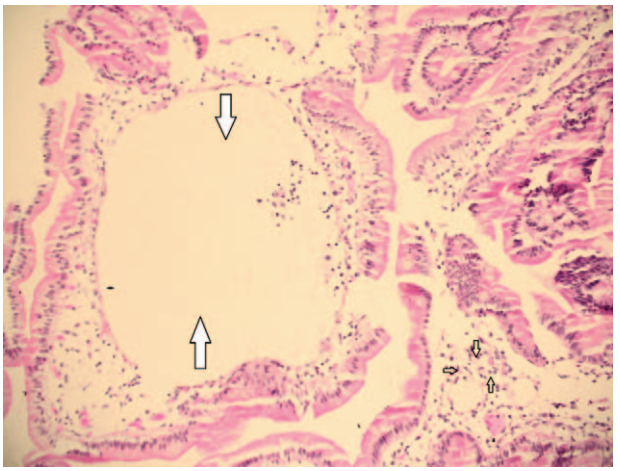

Figure 2: Duodenal biopsy showing dilated lymphatics within the lamina propria (white arrows) with the presence of mild mixed inflammatory cell infiltrations (small arrows).

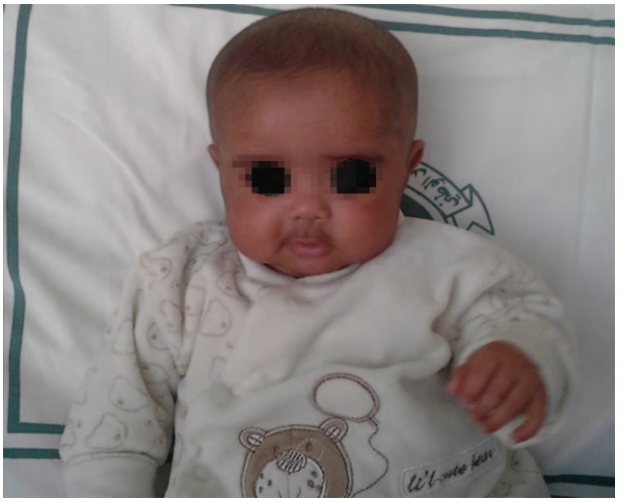

Figure 3: The same infant after Monogen formula and fat restriction (post-treatment).

Conflicts of interest

None.

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