Intestinal Lymphangiectasia

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Received: 11 July 2010
Accepted: 20 August 2010

History

A three year old boy presented with tetanus who additionally suffered from non-bloody diarrhea. Physical examination showed bilateral edema of the lower limbs.

Laboratory data showed iron deficiency anemia, hypoalbuminemia, hypocalcaemia and hypogammaglobulinemia. Stool culture was negative for bacteria however fat droplets were detected in the patient's stool. Endoscopic examination revealed white intestinal mucosa. Duodenal biopsy specimen showed dilated lymphatic channels in lamina propria (Fig. 1).

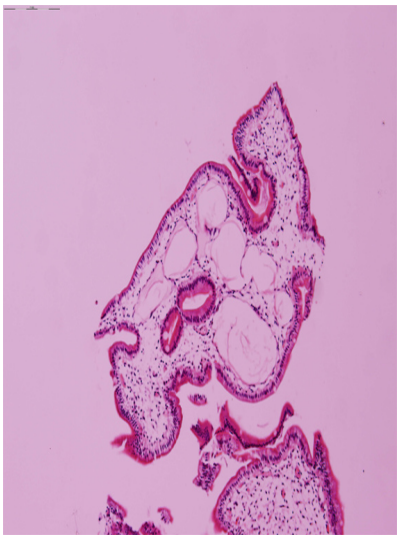

Fig 1: Duodenal biopsy specimen showed dilated lymphatic channels in lamina propria.

Diagnosis

Primary intestinal lymphangiectasia, first described by Waldmann et al. in 1961, is a rare disorder resulting from improperly formed lymphatics in which the lymph vessels supplying the mucosa become enlarged and obstructed. The disease is usually diagnosed before three years of age but may be diagnosed later. Several genes, such as VEGFR3, PROX1 and SOX18 are involved in the development of the lymphatic system. The enlarged and weak lymph vessels carry lymphatic fluid poorly and the lymphatic fluid leaks back into the intestine, preventing fat and proteins from being absorbed into the bloodstream. Low protein levels cause tissue edema.

Serous effusions are relatively common. The main symptom is diarrhea and the definitive diagnosis is based on biopsy results. Hypocalcemia is a common finding due to the loss of calcium binder proteins.

A low-fat, high-protein diet with medium chain triglyceride (MCT) supplements assist with symptom management. The need for dietary control seems to be permanent, be-

cause signs and symptoms will reappear after treatment withdrawal. Our patient's symptoms are controlled with supportive diet and he remains problem-free after about three years of follow up.

In the cases of seizure-like attacks, electrolyte disturbances and hypoalbuminemia, the possibility of Protein Losing Enteropathy exists.