CONGENITAL INTESTINAL LYMPHANGIECTASIA: A CASE STUDY

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ABSTRACT
Congenital intestinal lymphangiectasia is a rare disease which leads to protein losing enteropathy that usually affected children and young adults. Tortuous, dilated lymphatic vessels in the intestinal wall and mesenterium are typical features of the disease. Major symptoms include peripheral edema, mal absorption, diarrhoea, steatorrhea, and effusions. Specific diet and medication are required for disease control. Specific diet and medication are required for disease control.

Keywords: Ileal diseases, lymphangiectasis, intestinal, congenital abnormalities, diagnosis, differential, drug therapy.

CASE REPORT
A 20-year old student with no known comorbidities was presented in Gastro Enterology department of a tertiary care teaching hospital. On medical history taking it was revealed that 11 months back he had developed mild edema at both lower limbs and was increased progressively and exhibited at abdominal area too. He was consulted at a local hospital and was prescribed with anti-inflammatory and diuretics to reduce edema but there was no symptomatic relief. By the meantime he developed facial puffiness in the morning hours and had intermittent episodes of loose stools with presence of blood and mucus in the stool. Baseline investigation was found to have hypoprotenimia - 2.26 g/dl [reference range 6.6-8.3(g/dl)] and hypocalcenemia 4.4mg/dl [reference range 8.8-10.6 (mg/dl)]. He was started with high protein diet, spironolactone was given for calcium correction. Ascitic tapping was done and fluid was sent for detailed investigation, nature of ascitic fluid was milky. oesophago-gastro deuodenoscopy showed intestinal lymphangiectasia. HRCT chest was done to rule out secondary causes and was found to be
normal. Lymphoscintillography was done and findings are suggestive of mild to moderate leak into abdominal cavity probably leak from level of cisterna chyli. Enteroscopy demonstrated dilated lymphatic vessels in the small intestine. The diagnosis was confirmed by intestinal biopsy. The patient was put on high-protein diet containing medium-chain fatty acids along with supportive therapy. The discharge medication was T. Furosemide 40mg 1-0-0, T. Spironolactone 50mg 1-1-0, T. folic acid 5mg Od, T. Shelcal 500mg (Calcium Carbonate) bd).

DISCUSSION
The disease can be primary (congenital), where there is malformation within the lymphatic channels leading to their blockage, and the condition is usually diagnosed within the first decade of life. The condition can also be secondary to other disease states, such as constrictive pericarditis, lymphoma, sarcoidosis and scleroderma, and it can affect older adults. The first manifestations are usually persistent diarrhea and peripheral edema. The edema can be unilateral or bilateral, and macular edema revealed in funduscopic examination has been reported. The lymphatic hypoplasia results in an obstruction in lymph flow, which leads to increased pressure within the lymphatics. This, in turn, will cause dilation of the lymphatic channels in the intestine and, finally, lead to the rupture of the channels and resultant discharge of the lymph into the lumen of the bowel. The characteristic endoscopic and radiographic features of intestinal lymphangiectasia have been documented thereby greatly facilitating an accurate diagnosis.

In this case, the first presentation was found to be hypoproteinemia. After excluding synthetic obstacles and insufficient intake as the causes, we concluded that protein loss was the main factor. We also repeated some tests to exclude proteinuria. At last, we were sure that it was a protein-losing gastrointestinal disease.

CONCLUSION
Congenital intestinal lymphangiectasia is a rare disease, usually diagnosed in childhood. Early recognition of the disease and adequate treatment can prevent development of various complications. A positive clinical response to the special diet therapy, namely a low-fat and medium-chain triglyceride diet, can further confirm the diagnosis of primary intestinal lymphangiectasia.
REFERENCES


