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# Congenital Intestinal Lymphan- giectasia: Report of a Case

Congenital intestinal lymphangiectasia is a rare protein-losing enteropathy that usually affects children and young adults. Major symptoms include peripheral edema, mild non-bloody diarrhea, and chylous effusions that may develop during the course of the disease.

In this disorder intestinal lymphatic vessels show fibrous occlusions that lead to pressure elevation of the lymphatic flow and rupture of the small lymphatic vessels. Transudation of lymph fluid into the different layers of the intestinal wall and lumen then occurs.

**Keywords:** Congenital, Intestinal Lymphangiectasis, Lymphatic System

## Introduction

Congenital intestinal lymphangiectasia is characterized by hypoplastic lymphatic vessels in the mucosa and submucosa of the small bowel.[1-3] Following the obstruction of the lymphatic flow, dilation of lymphatic channels occurs. As a result, the bowel wall thickens due to edema and congestion.[2-5] Rupture of these dilated lymphatic vessels causes entrance of the lymph fluid into the different layers of the bowel wall and the lumen. Sometimes the lymph fluid enters into the peritoneal cavity and the pleural space, and causes chylous effusions. Hypoproteinemia, lymphocytopenia, and steatorrhea may occur.[1-5] Intermittent peripheral edema is another symptom, which may become persistent later. This disorder affects mainly children and young adults.[1-3] There is no exact information about the incidence and prevalence of this disorder, and there are some case reports. In this case we found different imaging findings in abdominal and pelvic sonography, CT scan, and small bowel barium study. These findings correlate with the pathology and other imaging signs that were reported in literatures. Some of these findings include mucosal thickening and the characteristic halo sign on CT scan. In this case we saw the halo sign in sonography that correlates with the CT scan finding. This ultrasound finding has not been reported up to now. Early diagnosis of this congenital disorder can help many patients, because a low-fat diet with medium-chain triglycerides (MCT) may result in significant improvement of hypoproteinemia and clinical symptoms.[2]

## Case Presentation

A 42-year-old man was admitted to the hospital because of weakness, malaise, abdominal discomfort, and swelling of the legs. He was a known case of lymphangiectasia that was proven by histopathology (Fig. 1).

His problem began since childhood. He noticed tiny nodules with spontaneous white discharge on the scrotum when he was about 8 years old. He had a long history of recurrent non-bloody diarrhea, intractable ascites and abdominal pain. The abdominal pain was colicky and did not relate to food intake. There was no major weight loss or reduced appetite.

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On physical examination, positive findings included non-pitting edema and elephantiasis appearance in the legs, especially the left side, lymphadenopathies with soft consistency in the inguinal regions, and ascites.

Different investigations were achieved to rule out the secondary type of lymphangiectasia.

Laboratory tests were negative except for the hypoproteinemia and lymphocytopenia. The liver function tests and bilirubin (direct and indirect) were within normal range. BUN, creatinine and urine analysis were normal. Ascitic fluid was chylous and was negative for bacteria, mycobacteria, and fungi. Bronchoscopy showed no evidence of tuberculosis or cancer.

Laparoscopy was carried out to detect the probable cause of chylous ascites. The liver, the round ligament, the surface of the bowel, and the abdominal wall were inspected. The round ligament was thick due to non-inflammatory edema. The liver seemed normal. There were fibrin like deposits on the surface of several segments of the bowel loops that were associated with lymphangiectasia. No evidence of tuberculosis or cancer was seen in the abdominal cavity.

Finally, no etiology was found and the diagnosis of primary (congenital) intestinal lymphangiectasia was confirmed.

The patient underwent different imaging investigations (sonography, barium study, and CT scan).

Barium study of the small intestine demonstrated moderately dilated small bowel loops and thickening of the mucosal folds. Flocculation due to hypersecretion, and the stacked-coin appearance was seen. Nodular pattern was detected that seemed to be associated with dilated lymphatics. Separation of the loops was due to the presence of ascitic fluid that is a common finding in this disorder. Opaque densities in the right upper quadrant on plain abdominal film, proved to be related to stones in the gall bladder at sonography (Fig. 2).

CT scans were performed on a GE 9800 high light advantage scanner. Pleural effusion was detected in both hemithoraces, especially the right side, which was septated in some locations. Ascitic fluid was discovered in the abdominal cavity. Gallstones were seen. No lymphadenopathy was detected in the abdominal cavity. Dilation and diffuse mural thickness of the bowel loops were observed. Hypodense bands throughout the mesentery seemed to be due to dilated lymphatic channels.

An important finding on CT scan of our patient was the presence of the halo sign that was due to the infiltration of the lymph fluid into the layers of the bowel wall and was consisted of three rings.

Other organs appeared normal in CT scan (Fig. 3).

In sonography of the lower zones of the abdomen, large cystic structures with fine septa, which seemed to be lymphoceles, extended to the pelvic floor (Fig.

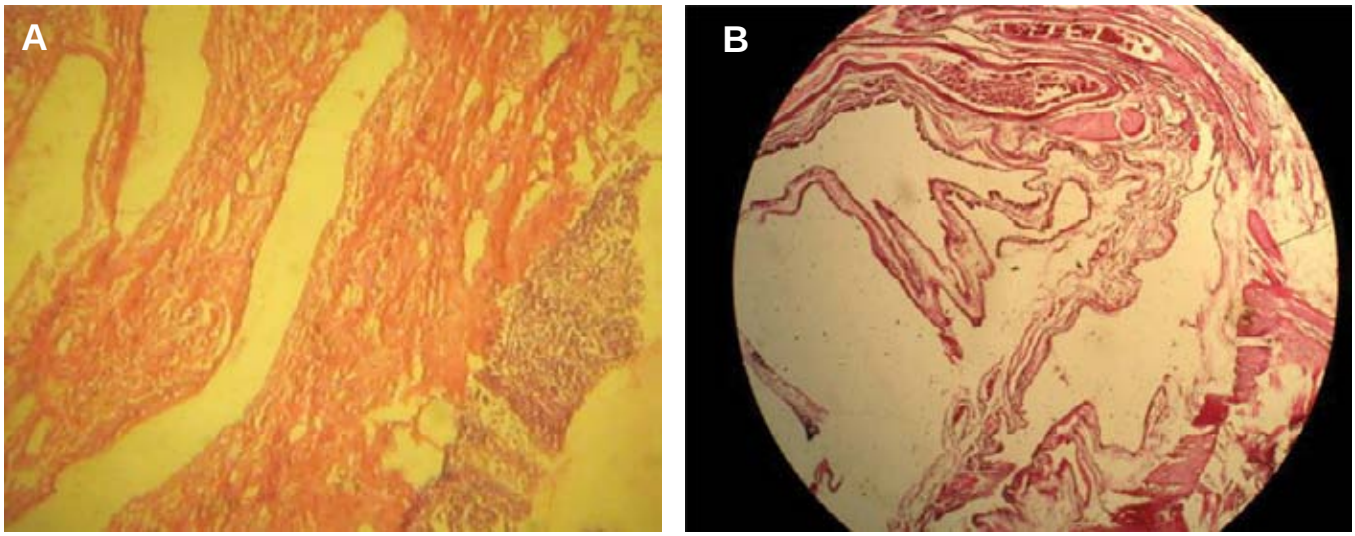


**Fig. 1.** 42-year-old man with congenital intestinal lymphangiectasia.

**A.** Biopsy of inguinal lymph nodes. It shows proliferation and dilation of lymphatic vessels with a thin wall in the connective tissue, the lymph node appears normal, which is compatible with lymphangioma.

**B.** Biopsy of the small intestine. Dilated lymphatics are compatible with intestinal lymphangiectasia.

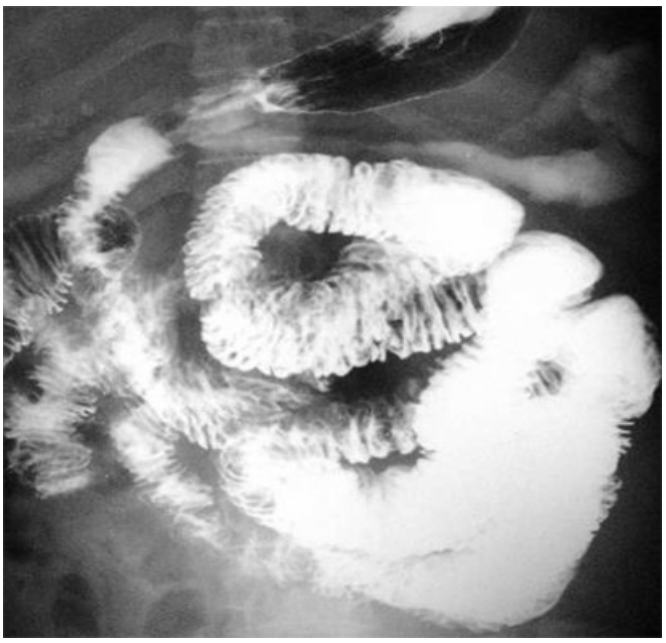

**Fig. 2.** Small bowel barium study shows moderately dilated loops and mucosal thickening with nodular pattern. Stacked coin appearance is seen. Opaque densities (gallstones) are seen in the right upper quadrant.

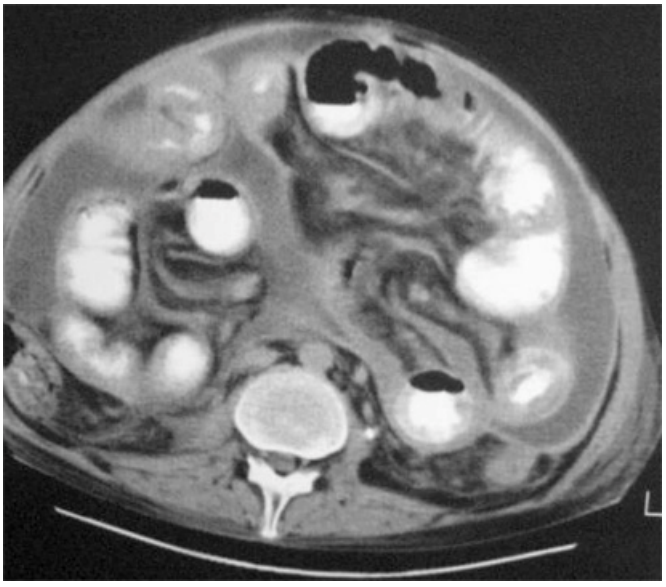

**Fig. 3.** Dilation, mural thickening of the small bowel and 'Halo' sign on CT scan. Ascitic fluid is detected.

4). Hydrocele was detected and we noticed ascitic fluid in the abdominal cavity.

Sonography revealed dilation of small bowel loops with increased mural thickness. The mucosa, submucosa and other layers of the bowel were seen with increased thickness and different echoes that mimic the halo sign on CT scan. We called it "*sonographic halo or target sign"* (Fig. 5).

Other organs seemed normal sonographically except for the presence of gallstones.

## Discussion

Intestinal lymphangiectasia is characterized by loss of protein and other lymph contents into the intestinal lumen.[2,5] This rare disorder can be primary (Milroy disease) or secondary. The primary type results from a congenital abnormality in the lymphatic system, and the secondary form may be due to surgical damages to the lymphatic vessels, chronic right ventricular failure, constrictive pericarditis, retroperitoneal tumors, tuberculosis and inflammatory bowel diseases.[3,5] Our patient underwent different investigations to rule out the secondary type. No etiology was found and the diagnosis of congenital intestinal lymphangiectasia was made.

Congenital intestinal lymphangiectasia usually presents in the childhood and early adolescence. Males and females are equally affected.[1,2] This disorder usually presents as a sporadic disease; however, there are several reports suggesting a genetic etiology in some cases.[3] This disorder manifests with episodic abdominal pain, steatorrhea, peripheral edema, and ascites.

Our patient was referred to the hospital with the same presentation and underwent different investigations.

Primary intestinal lymphangiectasia is usually limited to the small intestine, although colonic manife-

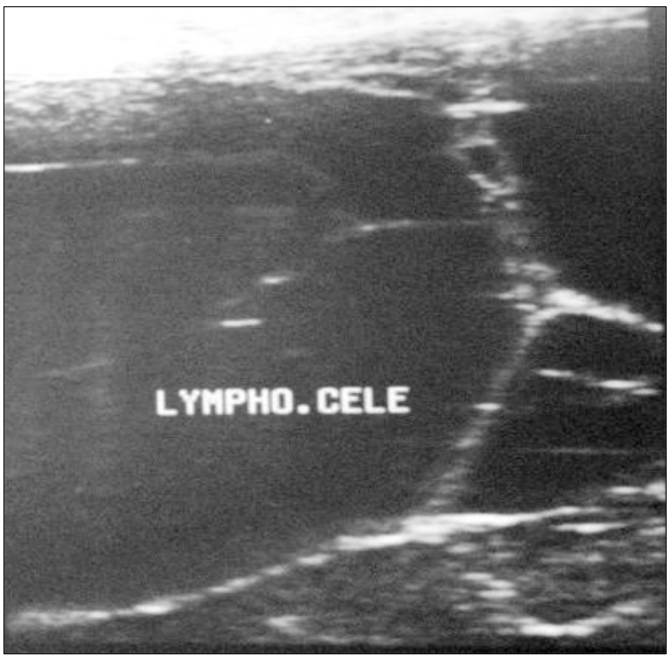


**Fig. 4** Lymphoceles with fine septations in the lower zone of the abdomen in abdominal sonography.

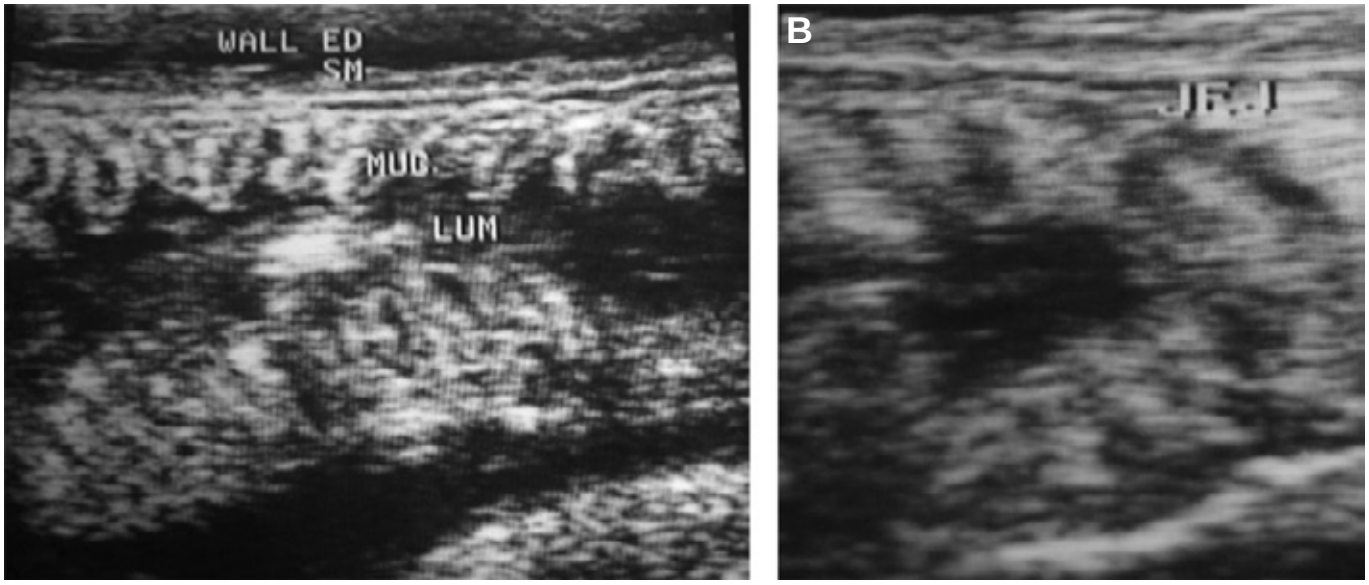


**Fig. 5.** Abdominal sonography in our patient with intestinal lymphangiectasia.

**A.** Longitudinal scan shows dilation and mural thickening of the bowel.

**B.** Transverse scan shows thickness of the mucosa, submucosa and other layers, which are edematous with different echoes and form the 'halo or target' sign.

stations have also been reported.[2]

Barium study of the small bowel demonstrates mild to moderately dilated loops and nodular mucosal pattern in more than 75% of the patients. The same pattern was seen in our patient. Separation of loops was due to chylous ascites, which is a common finding in this disorder. The sensitivity of this method, however, is not sufficient to determine the extraluminal and mesenteric abnormalities.[2]

Abdominal CT scan of such patients usually shows dilation and nodular mural thickening of the small bowel loops. Ascitic fluid is commonly seen. Hypodense bands in the small bowel wall and the mesentery due to dilated lymphatic channels are often seen. CT scan helps us to rule out the secondary causes of lymphangiectasia. An important CT scan finding in this disorder is the halo sign, which was first described in a patient with Crohn's disease. The halo sign consisted of two rings surrounding the bowel lumen. The inner ring with lower attenuation was suggestive of edematous and irregular mucosa, and occasionally submucosa. The outer ring with higher attenuation was associated with the other bowel layers.[6]

In primary intestinal lymphangiectasia, the halo sign is composed of three different rings. The inner ring with low attenuation is indicative of dilated lymphatic channels, focal edema, and an increased number of foamy histiocytes in the mucosal layer. The middle ring with very low attenuation is due to dilated lymphatic channels and submucosal edema. The outer ring has a higher attenuation and is related to the rest of the bowel wall (muscularis properia and serosa).[6] These findings were clearly seen in our patient.

Sonography shows diffuse mural thickening and dilation of the bowel loops and mesenteric edema, although it may be difficult to see the whole bowel lumen because of the intestinal gas.

We used a 7.5 MHz transducer to investigate the bowel loops and a 3.5 MHz transducer for solid abdominal organs. Increased thickness of the layers of the bowel wall with different echos was observed. Actually, we saw the sonographic halo or target sign, which was compatible with CT scan and histopathology. This sonographic sign has not been reported so far. We think that diffusion of lymph into the layers of the bowel wall results in thickening of the layers with different echoes, which forms the sonographic halo sign.

In this report, we studied a rare congenital enteropathy "primary intestinal lymphangiectasia", and could correlate imaging with histopathology. Of course, there are few similar cases with different degrees of the disorder. Involvement of the intestine causes similar patterns in barium study and CT scan

with intestinal dilation and mural thickening.[2-6] Some of these studies reported the halo sign in CT scan,[5,6] but non of them reported the sonographic halo sign as we mentioned in our case.

Our patient underwent a low fat regimen with medium chain triglycerides and other symptomatic therapies. He responded relatively well to this kind of therapy, and many of his symptoms like ascites and edema improved to some extent.

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