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Case Report

Small Bowel Lymphangiectasia Leading to Massive Gastrointestinal Bleeding: A Case Report

Farahnaz Safaei[1], Amir Sadeghi[1*], Pardis Ketabi Moghadam[1], Peyman Taheri[2]

[1]Gastroenterology and Liver Diseases Research Center, Research Institute for Gastroenterology and Liver Diseases, Shahid Beheshti University of Medical Sciences, Tehran, Iran
[2]Medical School, Mashhad University of Medical Sciences, Mashhad, Iran

Abstract

Lymphangiomas are benign lymphatic system abnormalities that can appear anywhere on the skin and mucous membranes. Lymphangiomas are caused by congenital or acquired lymphatic system disorders. In the congenital form, although the cause is unknown it is said that it is formed by the incorrect attachment of lymphatic channels to the main lymphatic drainage duct before the age of 5 years. lymphangiectasia as a subgroup of lymphangioma occurs seldom in the small bowel, especially in adults. If that happens, protein-losing enteropathy will be the most common presenting sign. In the present study, we introduce a case of a 40-year-old man without a history of any congenital or acquired diseases who was admitted to the emergency room due to long-lasting obscure overt gastrointestinal (GI) bleeding. Normal upper and lower GI endoscopies were suggestive of GI bleeding originating from the small intestine. Despite receiving iron supplements, he continued to have melena and remained anemic. Further evaluation of the small intestine by deep enteroscopy revealed multiple white spots histologically consistent with dilated lymphatics. Intestinal lymphangiectasia was eventually introduced to be the final diagnosis of the patient.

Keywords: Lymphangioma, Lymphangiectasia, Small bowel bleeding

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Introduction

Lymphangiomas are benign lymphatic system abnormalities that can appear anywhere on the skin and mucous membranes.[1] Lymphatic malformations are benign dilated or cystic lymphatic ducts and are classified into two groups of macrocystic (cystic hygroma) and microcystic (lymphangioma) groups. Cystic hygroma is usually congenital and is detected in the first trimester of pregnancy by ultrasonography and is usually related to chromosomal abnormalities. Lymphangiomas are made of intertwined threads of transparent or hemorrhagic lymphatic ducts, which are detected after birth or during the first years of life particularly involving skin or subcutaneous tissues.[2] The incorrect attachment of lymphatic channels to the main lymphatic drainage duct before the age of five has been known as an initiating factor for the congenital form.[3,4] Lymphangiectasia is a subcategory of lymphangioma that would be acquired or congenital. Intestinal lymphangiectasia has rarely been reported in the literature.[5] Any interruption of previously normal lymphatic drainage, such as surgery, cancer, radiation therapy, abdominal trauma, portal hypertension, tricuspid regurgitation, constrictive pericarditis, or lymphatic obstruction can result in acquired lymphangiectasia.[6,7] Its congenital form that is suggested to have an underlying genetic predisposition frequently has been reported in following syndromes like Turner, Yellow-nail syndrome, etc. in teenage.[5] Despite accounting for 90% of the mucosal surface area of the gastrointestinal (GI) tract, lymphangiomas occur seldom in the small bowel, especially in adults.[8-10] The most typical symptom of this condition is a non-tender, freely moving abdominal mass with a partial small intestine blockage. In these patients, intestinal obstruction may be caused by intestine adhesion due to an infected cyst or intestinal volvulus if they are close to the duodenojejunal junction.[9]

The presented case is a 40-year-old man without a history of any congenital or acquired diseases who was admitted to the emergency room due to long-lasting GI bleeding. Normal upper and lower GI endoscopies were suggestive of GI bleeding originating from the small intestine. Despite receiving iron supplements, he continued to have melena and remained anemic. Further evaluation of the small intestine by deep enteroscopy revealed multiple white spots histologically consistent with dilated lymphatics. Intestinal lymphangiectasia was eventually introduced to be the final diagnosis of the patient.

Case Report

We report a case of a 40-year-old man who was referred with a history of melena in the last 4 months leading to a significant drop in hemoglobin. Despite receiving 18 units of packed red blood cells (RBCs), the patient's

*Corresponding Author: Amir Sadeghi, Email: amirsadeghimd@yahoo.com

hemoglobin level was not corrected and was persistently 6 mg/dL during previous hospitalizations. When he was first brought to the emergency room in our center, his vital signs were stable. Physical examination showed pitting edema of the lower extremities on both sides, ascites, and decreased right lung sound indicative of pleural effusion. The patient had no history of medical problems, drug use, as well as tobacco, other illicit drugs, or alcohol abuse. The initial diagnosis was upper GI bleeding. On admission, intravenous fluid in addition to pantoprazole and octreotide drip were administered and the patient was hospitalized.

In the primary lab test, ferritin was 5 µg/L, hemoglobin was 6.2 mg/dL, and Albumin was 2.1 g/dL. Other parameters were within normal ranges. Endoscopy and colonoscopy were performed. The findings of the endoscopy were normal, just a few erosions were seen in the stomach and duodenum. In a colonoscopy, no obvious lesion was found but melena. To evaluate the small bowel as a probable source of bleeding, we proceeded with an abdomen and pelvic spiral computed tomography (CT) with intravenous (IV) contrast, which revealed multiple hypodense mesenteric lymphadenopathies along the small bowel mesentery with the largest one about 15×20 mm, edematous small bowel mesentery, and no para-aortic lymphadenopathy. Lymphoproliferative disorders of small bowel mesentery, celiac disease, or less likely collagen vascular disease or chronic infections of small bowel were considered at differentials respectively. Further evaluations like CT enterography and abdominal magnetic resonance (MR) angiography were comparable to the spiral CT scan and presented a marked wall and mucosal fold thickening of the jejunum, extensive nodular type increased density of the mesenteric fat, and multiple mild enlarged mesenteric lymph nodes. Portal veins, splenic veins, and superior mesenteric veins appeared patent without thrombosis or narrowing. Further evaluations for collagen vascular diseases and tumor markers were all negative. Then, because of iron deficiency anemia, abnormal imaging, and colicky abdominal pain enteroscopy was performed which showed diffuse white infiltration of the jejunum, and multiple biopsies were taken (Figure 1). Histological findings reported a significant dilation of the lymphatics in the mucosa extending into the submucosa. The intestinal epithelium appeared normal although occasional creamy yellow villi were seen more apparently on top of the villi all indicating the presence of submucosal lymphangioma. Microscopic view of jejunal biopsies revealed significant dilation of the lymphatics in the mucosa extending into the submucosal suggestive of intestinal lymphangiectasia (Figure 2).

Afterward, a laparoscopy of the small intestine was performed to investigate and rule out other probable differential diagnoses including lymphatic malformations and hemangiomas. Immunohistochemistry (IHC) study of the laparoscopic full-thickness small intestinal biopsy revealed: CD31: focal positive, CD34: positive,

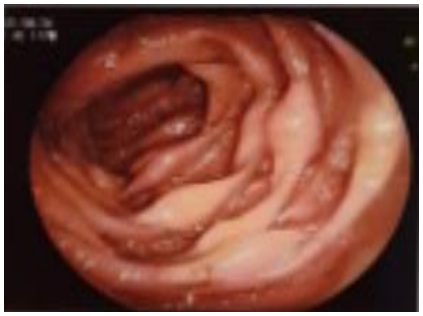

Figure 1. Infiltration of whitish spots was detected throughout the jejunum

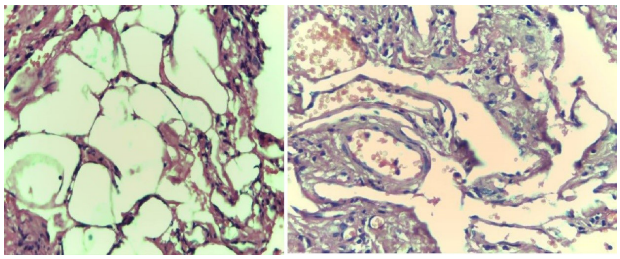

Figure 2. Microscopic view of jejunal biopsies

D2-40: positive in favor of lymphatic malformation (lymphangioma). To confirm the diagnosis, the patient's pathology slides were referred to the second pathologist who endorsed the mentioned diagnosis.

After lab tests and studies and a definitive diagnosis, the patient was discharged with medication instructions including esomeprazole 40 mg, subcutaneous octreotide, multivitamin-mineral supplement, dietary modification, and Ferinject ampoule. The patient is currently on the list for bowel transplantations in another center.

Discussion

Intestinal lymphangiectasia is reported to be unifocal, multifocal, or diffuse. Interference with the normal function of the lymphatic system would result in imbalanced tissue fluid, arresting the interstitial protein transport, interrupting immunological functions, and fat absorption. Symptoms associated with protein-losing enteropathy are commonly reported in the presence of lymphangiectasia.[11] Accordingly, lower limbs edema, pleural effusion, and ascites following hypoproteinemia and hypoalbuminemia in the absence of proteinuria seemed logical in the presented case although they were not being the case for referral to the hospital. When it comes to GI bleeding, a huge number of etiologies other than lymphangiectasia are prioritized in the differential diagnosis. Given the normal upper and lower GI endoscopies, we decided to proceed with diagnostic modalities for the evaluation of the small intestine. A spiral CT scan of the abdomen and pelvis with oral and intravenous contrast revealed multiple hypodense mesenteric lymphadenopathies throughout the small bowel mesentery in addition to small bowel wall enhancement, which raised suspicion of lymphoproliferative disorders. Additionally, celiac disease, collagen vascular disease, and

inflammatory or infectious disorders were differential diagnoses. These results were confirmed by CT-enterography and MR-angiography. The most reasonable approach to managing this patient with imaging harboring a malignant probability was balloon-assisted enteroscopy and deep mucosal biopsies. The endoscopic appearance of the small intestine manifested myriads of white punctate nodules and plaques throughout the duodenum, jejunum, and proximal parts of the ileum. Histopathological examination of taken specimens was compatible with lymphangiectasia, which was confirmed by IHC staining. Whether patient's massive GI bleeding could be attributable to intestinal lymphangiectasia or not was the outstanding question we have tried to answer. Per the revised guidelines, there are limited reports in line with intestinal lymphangiectasia as the final diagnosis of massive GI bleeding. Perisic and colleague introduced an 8-year-old girl with recurrent hematemesis and melena due to a focal duodenal lymphangiectasia without protein-losing-enteropathy signs and symptoms.[12] Another case of lymphangiectasia leading to protein-losing enteropathy and difficulty to control GI bleeding has been introduced by Gras et al in a 14-year-old boy after Fontan surgery.[13] It seems that an increase in the intra-lymphatic channels due to the obstacles which prevent normal chyle flow is the main culprit of opening the abnormal lymph-vessel connections leading to retrograde flow from the blood into the lymphatics and finally mucosal bleedings.[14,15] Park and colleagues also localized a small polypoid lesion pathologically consistent with lymphangiectasia responsible for melena in an 80-year-old woman, which responded to hot snare polypectomy of the lesion.[16] Another issue that was not answered in the case was the culprit abnormality leading to the massive lymphangiectasia at the age of 40. She was older than the one who had congenital lymphangiectasia and all secondary workups were negative to date for an acquired form of lymphangiectasia. Our patient's GI bleeding was controlled by conservative methods like dietary therapy with a high-protein, low fat, medium-chain triglyceride diet in addition to mineral and iron supplementation. Due to the extensive involvement of small intestine and the probability of rebleeding, he was also introduced and registered at Shiraz Transplant Center, located in Fars, Iran, for a probable small bowel transplantation.

Competing Interests

The authors declare no conflict of interest related to this work.

Ethical Approval

None.

References

1. Stewart CJ, Chan T, Platten M. Acquired lymphangiectasia ('lymphangioma circumscriptum') of the vulva: a report of eight cases. *Pathology* 2009;41(5):448-53. doi: 10.1080/00313020902885052
2. Martinez-Lopez A, Salvador-Rodriguez L, Montero-Vilchez T, Molina-Leyva A, Tercedor-Sanchez J, Arias-Santiago S. Vascular malformations syndromes: an update. *Curr Opin Pediatr* 2019;31(6):747-53. doi: 10.1097/mop.0000000000000812
3. Miceli A, Stewart KM. Lymphangioma. In: *StatPearls*. Treasure Island, FL: StatPearls Publishing; 2021.
4. Sehgal VN, Sharma S, Chatterjee K, Khurana A, Malhotra S. Unilateral, blaschkoid, large lymphangioma circumscriptum: micro- and macrocystic manifestations. *Skinmed* 2018;16(6):411-3.
5. Umar SB, DiBaise JK. Protein-losing enteropathy: case illustrations and clinical review. *Am J Gastroenterol* 2010;105(1):43-9. doi: 10.1038/ajg.2009.561
6. Jacobs JD, Swanson PE, Brentnall T. Protein losing enteropathy due to gut plasmacytoma polyposis. *Am J Gastroenterol* 2019;114(12):1834. doi: 10.14309/ajg.0000000000000291
7. Meadows J, Jenkins K. Protein-losing enteropathy: integrating a new disease paradigm into recommendations for prevention and treatment. *Cardiol Young* 2011;21(4):363-77. doi: 10.1017/s1047951111000102
8. de Latour RA, Kilaru SM, Gross SA. Management of small bowel polyps: a literature review. *Best Pract Res Clin Gastroenterol* 2017;31(4):401-8. doi: 10.1016/j.bpg.2017.06.003
9. Kumar B, Bhatnagar A, Upadhyaya VD, Gangopadhyay AN. Small intestinal lymphangioma presenting as an acute abdomen with relevant review of literature. *J Clin Diagn Res* 2017;11(6):PD01-PD02. doi: 10.7860/jcdr/2017/22703.9962
10. Erguney S, Teksoz S, Erdamar S, Ainechii S, Olgun DC. Extended pancreaticoduodenectomy for a huge cystic-cavernous lymphangioma: a case report. *JOP* 2012;13(3):289-91.
11. Jha AK, Thandassery RB, Goenka MK. Focal intestinal lymphangiectasia: an unusual cause of acute overt obscure gastrointestinal bleeding. *J Dig Endosc* 2014;5(4):162-3. doi: 10.4103/0976-5042.150668
12. Perisic VN, Kokai G. Bleeding from duodenal lymphangiectasia. *Arch Dis Child* 1991;66(1):153-4. doi: 10.1136/adc.66.1.153
13. Gras P, Gottrand F, Godart F. Gastrointestinal haemorrhage due to lymphangiectasia caused by protein-losing enteropathy in the Fontan circulation. *Cardiol Young* 2017;27(8):1641-3. doi: 10.1017/s1047951117000853
14. Davidson JD, Flynn EP, Kirkpatrick JB. Protein-losing enteropathy and intestinal bleeding. The role of lymphatic-venous connections. *Ann Intern Med* 1966;64(3):628-35. doi: 10.7326/0003-4819-64-3-628
15. Poirier VC, Alfidi RJ. Intestinal lymphangiectasia associated with fatal gastrointestinal bleeding. *Am J Dig Dis* 1973;18(1):54-8. doi: 10.1007/bf01072238
16. Park MS, Lee BJ, Gu DH, Pyo JH, Kim KJ, Lee YH, et al. Ileal polypoid lymphangiectasia bleeding diagnosed and treated by double balloon enteroscopy. *World J Gastroenterol* 2013;19(45):8440-4. doi: 10.3748/wjg.v19.i45.8440