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Case Report

Primary Intestinal Lymphangiectasia Diagnosed by Capsule Endoscopy

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Abstract

Primary intestinal lymphangiectasia (PIL) is a rare disorder in children and it is more exceptional in adults. PIL is caused by a diffuse or localized dilatation and/or rupture of intestinal lymphatic vessels in the mucosa, submucosa, or subserosa due to high pressure in lymphatic vessels. The diagnosis is made on clinical grounds with the support of small bowel biopsies. The following report present a case of intestinal lymphangiectasia revealed by capsule endoscopy examination. This work shows that standard EGD and colonoscopy may miss characteristic lesions of PIL, and capsule endoscopy (or enteroscopy) may be required for the diagnosis because lesions are typically located in distal duodenum/jejunoileum.

Keywords: Primary intestinal lymphangiectasia, Waldmann's disease, Protein-losing enteropathy, Capsule endoscopy, Liver enzymes elevation, Case report.

INTRODUCTION

Primary intestinal lymphangiectasia is a rare protein-losing enteropathy (PEL) and was first described by Waldmann T *et al*., in 1961 [1]. This disease is due to obstruction of the lymph drainage system. Elevated pressure of lymph drainage in the intestinal wall can lead to dilatation and even rupture of the lymphatic vessels which, in turn, results in the leakage of lymphatic fluid. Intermittent diarrhea and edema, accompanied by other signs of malnutrition and hypoalbuminemia, are the most common symptoms of PIL.

We present a case of PIL accompanied by hepatic aminotransferase elevation, and diagnosed by using videocapsule endoscopy.

CASE PRESENTATION

A 65-year-old woman had been suffering from a moderate oedema of the lower extremities with ascites and intermittent pleural effusion for almost 2 years. The patient's weight was consistent with 85 kg at a height of 155 cm [body mass index (BMI): 35.3 kg/m²]. Pulmonary and renal diagnostics showed no underlying disease. The blood tests showed lymphocytopenia, a protein deficiency with a chronic lowered total serum protein (45 g/l; normal range 64–83 g/l) and hypoalbuminemia (21 g/L). Prothrombin time was normal (100 %). She had no anemia or hypocalcemia. We evaluated intestinal protein loss by fecal alpha-1-antitrypsin clearance, which was found to be > 24 mL/24h.

Laboratory examinations displayed also increased amino transferase (less than twice the normal upper limit) and gamma-glutamyl transferase (7 times the upper limit of normal), leading us to exclude all known causes of chronic liver disease: chronic viral hepatitis; hemochromatosis and hemosiderosis; autoimmune hepatitis and primary biliary cholangitis. There was no history of alcohol or potential hepatotoxic drug use. Cardiac failure was ruled out by echocardiography.

Abdominal ultrasound and computed tomography (CT) scan detected a small amount of fluid in the Douglas space and right pleural effusion.

The liver biopsy found: nodular regenerative hyperplasia, peliosis, focal and obliterating sinusoidal dilatation of portal veinlets. There were no signs of hepatic steatosis or other liver diseases.

Gastroscopy and colonoscopy with biopsies gave no pathological findings. Further diagnostic steps including a capsule endoscopy revealed the PIL of the stomach duodenum and proximal jejunum (Fig 1).

The diagnosis of Waldmann's disease was retained because there was no evidence of secondary causes of intestinal lymphangiectasia.

A few days after her discharge the patient died at home from massive digestive bleeding (no post-mortem autopsy had been performed).

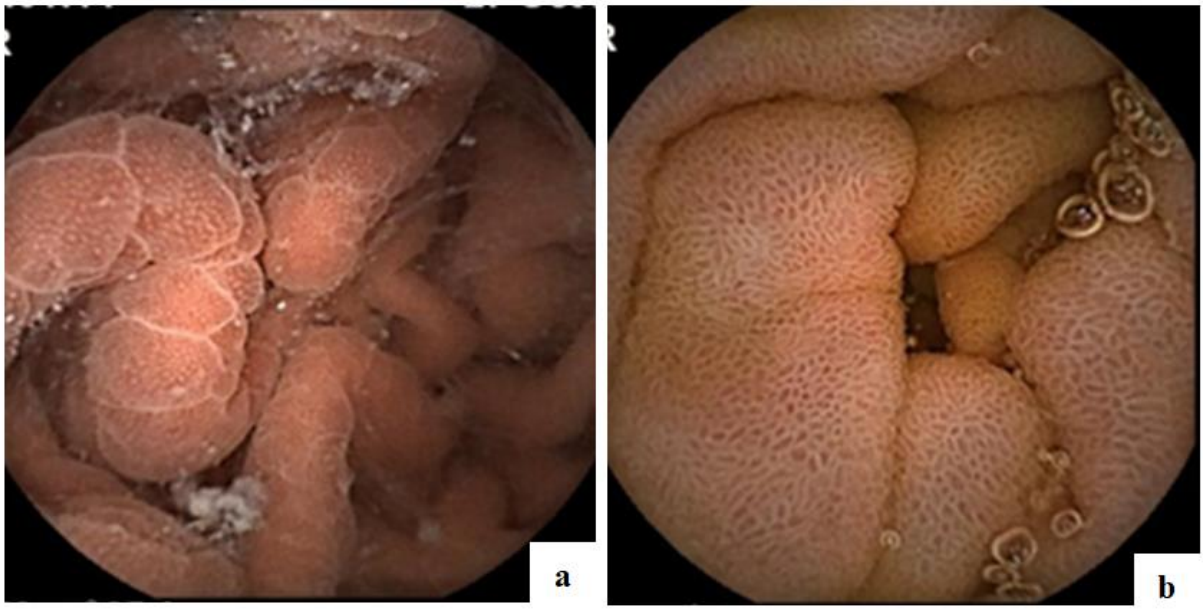

Figure 1: Capsule endoscopy findings; a) Diffuse edematous mucosae in the stomach with white curved lines probably related to submucosal dilated lymphatics and lacteal juice, b) Diffuse finger-like elongated mucosa covered with enlarged whitish villi in the duodenum

DISCUSSION

PIL is generally diagnosed before 3 years of age but it is not exclusively a childhood disease. PIL can be the sole finding in rare individuals or occur as part of a multisystemic genetic syndrome such as Hennekam syndrome, Turner syndrome, and Noonan syndrome [2].

As lymphatic fluid contains a lot of protein, fat and lymphocytes, leakage of lymph will cause hypoproteinemia, lymphocytopenia and decreased serum levels of immunoglobulin. Clinical manifestations may be presented as asymptomatic or fatigue, lower abdominal pain, edema, chylothorax, chronic diarrhea, steatorrhoea, ascites [3]. Iron deficiency anemia and hypocalcemic tetany can be observed too. Growth retardation is sometimes present. Lower gastrointestinal bleeding is rarely noted. It is secondary to small non-specific ulcers on the intestinal mucosa or a possible decrease in the coagulation factors [4].

The association between PIL and primary liver fibrosis is uncommon. Early detection of liver involvement in PIL is important in order to promote regression and prevent progression towards portal hypertension and recurrent cholangitis [5]. To the best of our knowledge, only six published cases reported the involvement of the liver in the PIL [5-8].

In our case, the liver biopsy found Peliosis hepatis (PH) signs. The pathogenesis of PH includes sinusoidal cell proliferation, which obstructs blood flow and causes pre-sinusoidal portal hypertension [9]. PH has been described in patients with conditions leading to advanced cachexia states (tuberculosis, AIDS, tumors) and in patients receiving certain drugs in particular anabolic steroids, oestroprogestogens, androgens and azathioprine. Patients with PH are often asymptomatic, but may develop progressive fibrosis, cirrhosis, and portal hypertension.

Diagnosis and treatment of PIL can be a challenge for clinicians because of the lack of specific guidelines. A diagnosis of PIL, by definition requires exclusion of secondary causes and it relied on specialized examinations, such as Push–pull enteroscopy with biopsy and capsule endoscopy of the small intestine. Whole-body MR imaging has significant advantages in comparison with other imaging modalities, noncontrast MR lymphography is useful for positive diagnosis, differential diagnosis, and specific evaluation of lymphedema severity [10].

PIL is often identified by upper gastrointestinal endoscopy and confirmed by pathology if the lesions involve the duodenum (a focal presentation is often seen in patients with acquired or secondary lymphangiectasia, in contrast to those with congenital disease who typically present with diffuse lymphangiectasia). The presence of patchy areas of lymphangiectasia in the jejunum and ileum cannot be detected because of the length limitation on upper gastrointestinal endoscopy. Capsule endoscopy is a

noninvasive and interesting tool to detect intestinal lymphangiectasia when other methods are incapable of finding the lesion [11]. Capsule endoscopy can also confirm the limited extension of PIL and serve as valuable preoperative tool.

Treatment of intestinal lymphangiectasia is primarily based on dietary therapy, with high levels of protein and low-fat intake supplemented with medium chain triglycerides (MCT). Surgery and drugs such as octreotide and sirolimus or everolimus are an alternative treatment for patients who are refractory to dietary therapy. lymphatic embolization could be a viable option when surgical resection or ligation is infeasible [12].

Long-term follow-up is needed since lymphoma have been described as long-term complications in patients with PIL [13].

CONCLUSION

PIL is an unusual cause of protein-losing enteropathy and it is a diagnosis of exclusion (secondary causes of intestinal lymphangiectasia should be excluded). Videocapsule is a useful tool to detect the presence of intestinal lymphangiectasia when the endoscopy is negative.

Conflict of Interest Statement: None declared.

REFERENCES

1. Waldmann, T., Steinfeld, J. L., Dutcher, T. F., Davidson, J. D., & Gordon Jr, R. S. (1961). The role of the gastrointestinal system in “idiopathic hypoproteinemia”. *Gastroenterology*, 41(3), 197-207.
2. Wang, N., Shi, W., & Jiao, Y. (2020). A PTPN11 mutation in a woman with Noonan syndrome and protein-losing enteropathy. *BMC gastroenterology*, *20*(1), 34.
3. Huber, R., Semmler, G., Mayr, A., Offner, F., & Datz, C. (2020). Primary intestinal lymphangiectasia in an adult patient: A case report and review of literature. *World journal of gastroenterology*, 26(48), 7707–7718.
4. Maamer, A. B., Baazaoui, J., Zaafouri, H., Soualah, W., & Cherif, A. (2012). Primary intestinal lymphangiectasia or Waldmann's disease: a rare cause of lower gastrointestinal bleeding. *Arab journal of gastroenterology : the official publication of the Pan-Arab Association of Gastroenterology*, *13*(2), 97–98.
5. Licinio, R., Principi, M., Ierardi, E., & Di Leo, A. (2014). Liver fibrosis in primary intestinal lymphangiectasia: An undervalued topic. *World journal of hepatology*, 6(9), 685–687.
6. Milazzo, L., Peri, A. M., Lodi, L., Gubertini, G., Ridolfo, A. L., & Antinori, S. (2014). Intestinal lymphangiectasia and reversible high liver stiffness. *Hepatology (Baltimore, Md.)*, 60(2), 759–761.
7. Jain, A. K., Agrawal, R., Hirani, S., Sircar, S., & Jain, S. (2020). A Family with Primary Intestinal Lymphangiectasia and Its Association with Liver Fibrosis. *J Liver Disease Transplant*, 9, 4.
8. Nguyen, C. D., Buchman, A., Bui, H. H., Thi-Lan Pham, A., Thi-Ngoc Vo, D., Dang, L. M., & Vo, V. H. (2022). Adult-Onset Primary Intestinal Lymphangiectasia With Liver Enzymes Elevation. *ACG case reports journal*, *9*(12), e00939.
9. Tsokos, M., & Erbersdobler, A. (2005). Pathology of peliosis. *Forensic science international*, 149(1), 25–33.
10. Arrivé, L., Derhy, S., El Mouhadi, S., Monnier-Cholley, L., Menu, Y., & Becker, C. (2016). Noncontrast Magnetic Resonance Lymphography. *Journal of reconstructive microsurgery*, 32(1), 80–86.
11. Wu, J., Huang, Z., Ji, M., Jiang, Z., Wang, Y., Tang, Z., & Huang, Y. (2021). The diagnostic value of capsule endoscopy in children with intestinal lymphangiectasia. Revista espanola de enfermedades digestivas: organo oficial de la Sociedad Espanola de Patologia Digestiva, 113(11), 765–769.
12. Kwon, Y., Kim, E. S., Choe, Y. H., Hyun, D., & Kim, M. J. (2021). Therapeutic Lymphatic Embolization in Pediatric Primary Intestinal Lymphangiectasia. *Yonsei medical journal*, 62(5), 470–473.
13. Huber, R., Semmler, G., Mayr, A., Offner, F., & Datz, C. (2020). Primary intestinal lymphangiectasia in an adult patient: A case report and review of literature. *World journal of gastroenterology*, 26(48), 7707–7718.