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CASE REPORT

A primary intestinal lymphangiectasia patient diagnosed by capsule endoscopy and confirmed at surgery: A case report

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Abstract

Intestinal lymphangiectasia (IL) is a rare disease characterized by dilated lymphatic vessles in the intestinal wall and small bowel mesentery which induce loss of protein and lymphocytes into bowel lumen. Because it most often occurs in the intestine and cannot be detected by upper gastroendoscopy or colonoscopy, and the value of common image examinations such as X-ray and computerized tomography (CT) are limited, the diagnosis of IL is difficult, usually needing the help of surgery. Capsule endoscopy is useful in diagnosing intestinal diseases, such as IL. We here report a case of IL in a female patient who was admitted for the complaint of recurrent edema accompanied with diarrhea and abdominal pain over the last twenty years, and aggravated ten days ago. She was diagnosed by M2A capsule endoscopy as a primary IL and confirmed by surgical and pathological examination.

Key words: Intestinal lymphangiectasia; Capsule endoscopy; Hypoproteinemia; Lymphocytopenia; Edema; Surgery

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INTRODUCTION

Intestinal lymphangiectasia (IL) is a rare disease and was first reported by Waldmanin in 1961[1]. Since then, no more than two hundred cases have been reported. IL can be classified into primary (idiopathic) and secondary type. Secondary IL is manifested as hypoproteinemia, lymphocytopenia and edema. IL has been found to be the cause of one third of protein-losing enteropathy patients. Main laboratory findings of this disease include decreased lymphocytes, serum protein and immunoglobin, increased fat concentration in stool. Common image examinations, such as lymphangiography, ECT examination, abdominal CT scan, are helpful. Endoscopy and capsule endoscopy can discover lesions which cannot be detected by gastroendoscopy or colonoscopy. Pathological examination makes its definite diagnosis. Medium chain triglyceride (MCT) supplement can improve its clinical symptoms. It was reported that somatostatin and antiplasmin are effective in some cases[2]. Surgery can be performed to alleviate symptoms in patients with limited lesions or a large amount of ascites[3]. Treatment of secondary IL is focused on symptoms and etiology. IL can progress to lymphoma after years or decades. We here present a case of IL diagnosed by capsule endoscopy and confirmed at surgery.

CASE REPORT

A 55-year old female was admitted to our hospital in March 2005 for recurrent edema accompanied with diarrhea and abdominal pain during the last twenty years, and aggravated ten days ago. The clinical findings are listed in Table 1. The patient denied other systemic diseases and any surgical history, as well as family history of such a disease. Other examinations (such as stool and urea examinations, serum tumor markers, purified protein derivatives (PPD) test, autoimmunity examinations) were all negative. Finally the patient underwent surgical exploration with resection of the affected intestine. Pathological examination confirmed the diagnosis of primary intestinal lymphangiectasia.

DISCUSSION

IL is a rare disease characterized by dilated lymphatic vessles in the intestinal wall and small bowel mesentery which induce loss of protein and lymphocytes into bowel lumen. This disease is due to obstruction of the lymph drainage system with various causes. When the pressure of lymphatic vessels increases, the lymphatic vessels dilate and rupture, resulting in leakage of lymph fluid, which in turn, leads to hypoproteinemia, edema, and lymphocytopenia.

Table 1 Clinical manifestations of the patient

Main clinical manifestations	Chronic diarrhea, lower limb edema, upper abdominal pain.
Laboratory possitive findings	Lymphocytes: 1.0 × 10^3/dL, albumin: (19.5-24.5) g/L, IgG: 407.0 mg/dL, IgM: 40.4 mg/dL, TSH (RIA): 5.58 mIU/L, freeT_3 (RIA): 1.24 pmol/L, freeT_4 (RIA): 4.54 pmol/L.
Endoscopy findings	Basically normal
Image findings	Abdominal computer tomography (CT) (Figure 1) showing thickening of sinus ventriculi and jejunal mucosa, and the sign of chronic cholecystitis; ECT of lymphatic system showing no obviously abnormal.
Capsule endoscopy fingding	Diffuse dilatation of lymphatic vessels (Figure 2A and B), thickening of intestinal villi. The dilatation of lymphatic vessels was more obvious in the middle and lower parts of the intestine. The dilated lymphatic vessels presented as coral and were mixed with the dilated capillaries.
Operation finding	Adhesion of greater omentum and abdominal wall. Gallbladder was enlarged and had multiple cholecystolithiasis, diffuse intestinal lesions and obviously dilated lymphatic vessels in the intestinal wall, most severe at the end of ileum.
Pathological findings	Dilated lymphatic vessels in the villi, submucosal and outside of the intestinal wall.

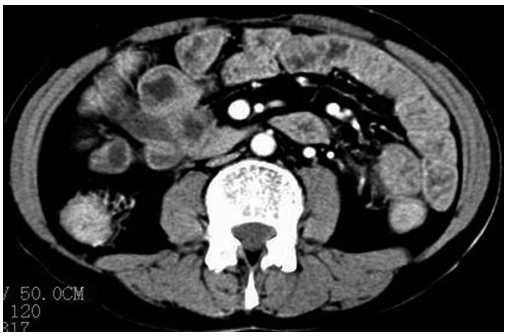

Figure 1 Abdominal computer tomography (CT) showing thickening of jejunal mucosa.

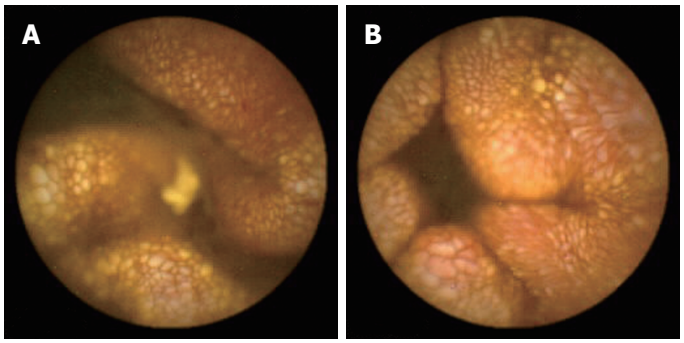

Figure 2 Capsule endoscopy showing diffuse oedematous aspect, dilatation of lymphatic vessels in the intestine (**A**) and thickening of whitish intestinal villi (**B**). The dilated lymphatic vessels presented as coral and were mixed with dilated capillaries.

Leakage of lymph fluid into the bowel lumen can result in celiac and steatorrhea and leakage of lymph fluid into the abdominal cavity can cause chyliform ascites. According to the cause of the disease, it can be classified into primary and secondary intestinal lymphangiectasia. Primary intestinal lymphangiectasia usually occurs in children and adolescents, due to deformity of the lymphatic system[1]. Adult intestinal lymphangiectasia is often secondary to many other factors, such as lymphoma, systemic lupus erythematosus (SLE), intestinal tuberculosis, retroperitoneal fibrosis, post-radiation effect[4], repeated parasite infection[5], hepatic cirrhosis[6], constrictive pericarditis, and abdominal surgery.

The disease is characterized by hypoproteinemia, lymphocytopenia, edema and ascites. Clinical manifestations may be presented as asymptomatic or fatigue, lower abdominal pain, edema, chylothorax, chronic diarrhea, ascites, recurrent intestinal bleeding with unknown reason, or protein-losing enteropathy, even as a surgical emergency with right abdominal pain, abdominal mass. Children may show growth retardation, smaller physical constitution. Iron deficiency anemia and hypocalcemic tetany can be observed too. Laboratory findings include decreased serum total protein and albumin along with decreased immunoglobulin levels including IgA, IgM, and especially IgG, due to its long half life. The number of lymphocytes, especially CD4+ and CD8+ lymphocytes, is decreased in almost every patient[7]. Secondary IL has other laboratory findings according to the primary disease. Diagnosis of this disease depends on characteristic features identified from gastroendoscopy and capsule endoscopy. Lymphangiography sometimes can find deformities of lymphatic vessels. ECT can indicate protein loss into the bowel lumen. Pathological examination confirms the final diagnosis. Since intestinal lymphangiectasia often occurs at jejunoileum[1,8], the upper and lower gastroendoscopy cannot access it in the intestine. If the lesion is within the reach of the endoscope, a biopsy would be diagnostic. Otherwise, lymphangiography or surgery is needed for a diagnosis. Capsule endoscopy is a relatively new technique, it is highly useful for evaluating small bowel diseases such as small bowel Crohn's disease[9]. The manifestations of intestinal lymphangiectasia observed by capsule endoscopy have been reported[10]. Capsule endoscopy is a useful and noninvasive method to diagnose intestinal lymphangiectasia when other methods are incapable of finding the lesion. Treatment of intestinal lymphangiectasia depends on different situations. Usually a low-fat, high-protein diet supplemented with medium-chain triglyceride (MCT) can improve the clinical symptoms and signs[11-13]. Essential trace elements need to be supplemented too. Both primary and secondary intestinal lymphangiectasia can follow this treatment with few side effects. However, patients may need to be treated in their whole life, because the symptoms and signs will recur soon after stopping the treatment. Medium-chain triglycerides, unlike the long-chain triglycerides, can bypass the lymphatic system and directly secreted into portal system, avoiding the increase

of lymphatic vessel pressure and the loss of protein. It was reported that somatostatins such as octreotide can decrease triglyceride absorption[6,14]. Antiplasmin therapy may have some role when fibrinolysis is increased[15,16]. Isolated lesions should be resected.

In this paper, we report a case of IL, mainly presented as protein-losing enteropathy. Capsule endoscopy is a good option for the diagnosis of intestinal diseases such IL.

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