CASE REPORT

An unusual case of chylous ascites: A case report

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Received: May 26, 2014 **Accepted:** July 1, 2014 **Online Published:** July 16, 2014
DOI: 10.5430/crim.v1n2p210 **URL:** http://dx.doi.org/10.5430/crim.v1n2p210

Abstract

Intestinal lymhangiectasia (IL) is a rare cause of intractable, but potential curable ascites. It is characterised by a dilation of the lymphatics of the mucosa, submucosa, or subserosa in the small intestine and subsequent loss of lymph fluid. The symptoms are edema, diarrhea, and chylous ascites. It has been considered a rare disease in children with congenital malformations, but has recently been diagnosed more frequently in adults. The latter is more often caused by a local obstruction of the lymph vessels. We report a case of primary IL in a 62-year-old man, who presented with chylous ascites, diarrhea, and edema. The symptoms dramatically improved with the recommended low-fat, protein-rich diet, supplemented with medium-chain triglycerides (MCT) and parenteral nutrition. After cessation of the parenteral nutrition the chylous ascites relapsed and the parenteral nutrition, MCTs and diet were reinitiated, but he did not need paracentesis anymore.

IL is now more frequently diagnosed, including asymptomatic cases being examined for other diseases. Therefore, it is important to be aware of this condition even in middle-aged patients since it can be treated and occationally cured.

Keywords

Intestinal lymphangiectasia, Chylous ascites, Diarrhea, Malabsorption, Edema, Hypoalbuminemia, Pleural effusion

1 Introduction

Ascites is a frequent complication to cirrhosis, peritoneal malignancy, right-sided heart insufficiency, and peritoneal tuberculosis. Occasionally, ascites occurs in patients with intestinal lymphangiectasia (IL) [1]. This condition is characterized by dilated lymphatic vessels in the mucosa, submucosa, or subserosa in the small intestines [1, 2]. Depending on the site of the lesions, lymph fluid drains into the intestinal lumen or causes formation of abdominal chylous ascites [2].

Typically, the patients develop hypoproteinemia, hypogammaglubulinemia, and hypoalbuminemia and subsequent fluid retention with peripheral edema, pleural- and pericardial effusions, and ascites [3]. Other symptoms include abdominal pain and diarrhea with malabsorption [1]. IL is most often caused by a congenital malformation of the lymph vessels and therefore diagnosed in childhood, but occasionally it presents with a late onset due to secondary obstruction of the lymph

vessels [3]. IL was first described in 1961 by Waldmann et al. and since then about 200 cases have been reported [1, 4-7]. We present a patient with primary IL as a rare cause of intractable, but potential curable ascites.

2 Case report

A 62-year-old male patient was referred to our department because of progressive formation of ascites. Over a period of six months the patient had developed peripheral edema, abdominal distension and an unintended weight-loss of approximately 20 kg. He had no history of contagious liver disease or ischemic heart disease. He had observed an intermittent diarrhea and constipation and had occasionally experienced blood in the stools. The fluid retention had been treated with furosemide and spironolactone without effect. Clinically, the patient was malnourished and had signs of distended abdominal veins and hepatic decompensation with ascites and peripheral- and scrotal oedema. However, the patient was without other cirrhotic stigmata.

A CT scan showed ascites and bilateral pleural exudates, but all other organs were normal. The hepatic vasculature was normal and the liver parenchyma was homogenous with a few liver cysts and opacities, that were identified as hemangiomas on ultrasound, but otherwise the morphology and size of the liver was normal.

The biochemistry revealed marginal unspecific anemia, normal levels of thrombocytes and leucocytes. CRP was 52 mg/L (normal reference level: 0-10 mg/L). Plasma creatinine and blood urea nitrogen were within the normal reference level. Serum albumin was reduced to 29 g/L, but all other biochemical markers of the liver were within the normal reference level, including INR, alkaline phosphates, aminotranferases, and α-fetoprotein. Blood tests for hepatitis B and C virus were negative.

Paracentesis revealed a large volume of chylous ascites. Analysis of the ascitic fluid showed low concentrations of amylase and LDH, a normal total protein level of 39 g/L and negative cultures. Cytological analyses of the ascitic and pleural fluids revealed no presence of malignant cells or Mycobacterium Tuberculosis, the quantiferon test was negative and CA19-9 was normal. The pleural fluid was described as chylous.

During hospitalization the patient continued to loose weight and suffered from alternating diarrhoea and constipation. Faeces blood test was positive, but levels of faeces calprotectin and elastase were normal. A colonoscopy revealed several benign polyps that were resected. Endoscopy of the esophagus and stomach did not show varices, but identified a polyp in the duodenum, which was characterised as gastric metaplasia at subsequent histological examination.

A lymph scintigraphy showed normal lymphatic flow from the lower extremities (see Figure 1). A 18F-PET-CT scan was without signs of cancer and cardiac function as assessed by echocardiography was normal. A liver vein catherization revealed a normal hepatic venous pressure gradient, ruling out postsinusoidal portal hypertension. A magnetic resonance imaging of the small intestines showed signs of intestinal wall thickening. A following diagnostic laparoscopy revealed edema and hyperemia in the proximal part of the small intestines. Double balloon enteroscopy showed a diffuse edematous mucosa in the small intestines, covered with enlarged and swollen villi containing whitish material (see Figure 2a, 2b and 3).

The patient continued to loose weight and was put on a protein-rich, low-fat diet, supplemented with medium-chain triglycerides (MCT), which was initiated together with full parenteral nutrition and octreotide 100 mg 3 times a day for 23 days. Treatment with octreotide had no substantial additional effect.

This diet resulted in a small weight gain, decreased the production of chylous ascites and the hemoglobin- and albumin levels increased. After the initial amelioration the improvement stagnated and for about six months he was without the need for paracenthesis.

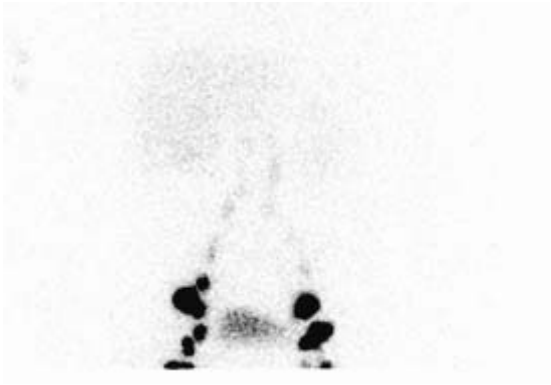

Figure 1. Lymph scintigram from the patient showing normal drainage from the lower extremities.

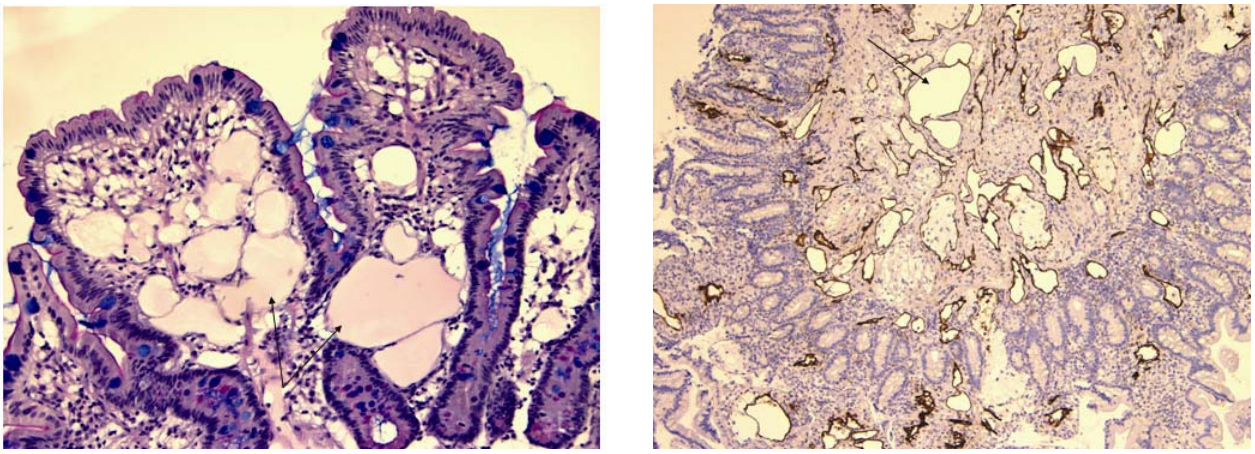

Figure 2. Biopsies from the small intestine showing dilated lymphatics in the lamina propria and submucosa compatible with intestinal lymphangiectasia. Panel a (left): Arrow showing dilated lymphatic spaces. Panel b (right): Immunohistochemical monoclonal antibody shows the lymphatic endothelium of dilated lymphatic spaces, also in submucosa. Arrow showing dilated lymphatics.

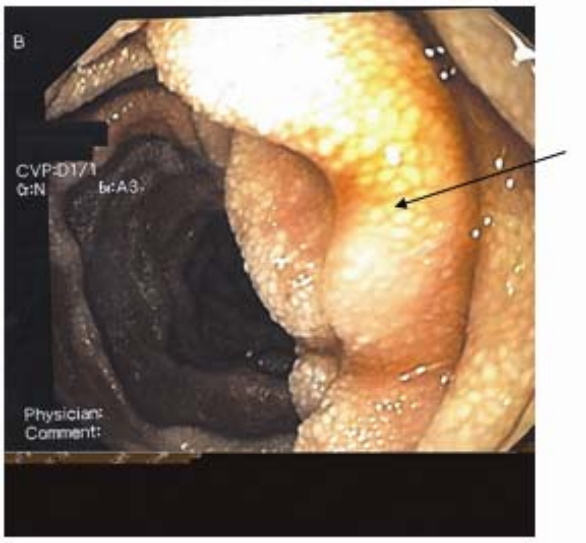

Figure 3. Jejunal mucosa as visualized by double-baloon enteroscopy, demonstrating white-tipped villi characteristic of intestinal lymphangiectasia. The arrow shows the white-tipped villi in the mucosa.

Suddenly, after six months the patient started vomiting and had difficulties swallowing food. A new 18F -PET-CT scan revealed a tumour in the transverse colon. Histologically, the tumour was an adenocarcinoma, pT2N0M0 and the patient was not offered chemotherapy. The patient underwent resection of the tumour and the omentum was described as inflamed but without any signs of lymph obstruction around the tumour.

There was no sign of metastases nine months after the operation.

Six months after the operation the parenteral nutrition was briefly discontinued, which resulted in increased production of the chylous ascites and the treatment was reinitiated. However, there has been no need of further paracentesis and the patient has a stable weight on the parenteral nutrition, MCTs and the protein-rich, low-fat diet.

3 Discussion

This case report presents a patient hospitalised because of chylous ascites, edema and diarrhea. The diagnosis was confirmed as IL. Two years after he presented with the first symptoms of IL a cancer was diagnosed in the colon. The patient had no metastases, the tumour was resected and the patient kept a stable weight on the protein-rich, low-fat diet, MCTs and parental nutrition and had no need of paracentesis anymore.

Previous case reports describe a great diversity in the course of the development of symptoms in patients with IL. Some patients have a short period of fever initially, while others present with acute ileus and some develop the symptoms gradually [2, 4, 5, 8-15].

The disease is most often seen in children, but in recent years it has been diagnosed more frequently in adults. IL can be classified as primary owing to congenital disorders or secondary as a result of other diseases that obstruct the lymph vessels. The latter can be caused by numerous conditions as for example cancer, rheumatic diseases, tuberculosis, Budd-Chiari syndrome, congestive heart failure, portal hypertension, chemotherapy, radiotherapy and other gastro-intestinal diseases, or surgery (see Table 1) [1, 8, 16, 17]. The diagnosis is based on endoscopic changes in the small intestine, and a histological picture that is dominated by dilated intestinal lacteals in the lamina propria of the villous structure. Occasionally the changes extend into the submucosa or even in the mesentery alone with minimal changes in the lamina propria [17]. The changes are most often observed in the duodenum and can be either segmental, multifocal or diffuse [1, 16].

Table 1. Etiologies of primary and secondary intestinal lymphangiectasia (IL).

Primary IL	Secondary IL
Congenital malformations incl. Turners syndrome	Cancer, lymphoma
	Rheumatic diseases, systemic lupus erythematosus, sarcoidosis
	Tuberculosis, pancreatitis, parasites
	Budd-Chiari syndrome
	Congestive heart failure
	Portal hypertension
	Retroperitoneal pathology or fibrosis
	Chemotherapy, radiotherapy, surgery

Studies have reported that several genes and regulatory molecules for lymphangiogenesis are related to primary IL [1, 13].

The clinical manifestations diverge from the patient being almost asymptomatic to show signs of malabsorption with malnutrition. This makes a true estimate of the incidence difficult to assess [10]. However, IL is now more often discovered incidentally in patients examined for other causes, and it seems to be more common than previously assumed [1, 13].

Due to the late onset of symptoms of IL in our patient we anticipated that he most likely suffered from secondary IL, but we were not able to find evidence to support this theory, since no of the known precipitating factors were identified. For this reason we consider our patient to be a case of primary IL [3].

Both primary and secondary IL has been associated with cancer. 5% of the patients with primary IL seem to develop lymphomas, but it is most often with a delayed onset of 31 years (range 19-45 years) after the initial diagnosis of IL.

In our case it is not likely that the adenocarcinoma was a result of the IL, since the most frequent malignancy associated with IL is lymphoma, which develops after decades. Moreover, adenocarcinoma in the colon is the third most common cancer in the western world today [18].

We find it less likely that the adenocarcinoma caused the IL. First of all the patient was diagnosed with gastrointestinal cancer almost two years after the first symptoms of the IL, a 18F- PET-CT scan 24 months prior to his cancer diagnosis, when the symptoms of IL developed, was without suspicion of malignancy and the colonoscopy performed 23 month prior to the cancer diagnosis was without suspicion hereof. Furthermore, the location of the cancer in the transverse colon can hardly explain the local lymphangiectasia in the small intestines and 6 months after the tumour was resected cessation of the parenteral nutrition resulted in increased production of chylous ascites.

The primary form of IL can be treated with a protein-rich, low-fat diet, supplemented with MCT, and in the case of the secondary form of IL, the underlying reason should be treated first [6, 16].

MCTs passes directly from the intestine to the portal venous system, rather than via the lymphatics, which reduces the risk of rupture by bypassing the engorged lymphatic vessels [6]. Treatment with anti-plasmin, steroids and octreotide has been tried, but their efficacies are variable reported and often insufficient [3, 19]. However, treatment with these drugs should be considered if the symptoms progress despite the dietary therapy.

Octreotide is a synthetic analogue of the naturally occurring hormone somatostatin, which is a potent inhibitor of the release of growth hormone, serotonin, gastrin, glucagon, and insulin. The mechanism is not well known, but is believed to involve reduction in lymph flow and immunomodulatory actions [20]. Surgery can be a successful treatment when fibrotic changes of the small bowel cause obstruction, but it is most desirable when the affected areas are local and not segmental [1, 21]. Even anastomosis of mesenteric to para-aortic lymph nodes has been tried [17].

In conclusion, IL is rarely diagnosed, but seems now to be more frequently revealed since some asymptomatic patients are diagnosed during examination for other disease causes. This case report emphasizes the importance of being aware of the condition even in middle-aged patients.

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