Chronic diarrhoea and abdominal mass: a case of intestinal lymphangiectasia

FM Trovato1, G Musumeci2*, G Bonanno3, C Pirri1, D Catalano1

Abstract

Introduction

Intestinal lymphangiectasia is a protein-losing enteropathy caused by congenital malformation or secondary obstruction of intestinal lymphatic vessels, which usually causes excessive protein loss in the intestine and malabsorption syndrome. Patients may be asymptomatic or complain of oedema, diarrhoea, hypoalbuminaemia and other deficiencies. This article reports a case of chronic diarrhoea and an abdominal mass.

Case report

We present the case of a 70-year-old woman who came to our gastroenterological clinic for diarrhoea. Blood and stool tests were within the normal ranges, except for a mild eosinophilia and a high C-reactive protein. The ultrasound scan showed a cystic retropancreatic mass, and the following computed tomography scan confirmed the presence of the mass without sure continuity with the dorsal pancreas. The fine needle aspiration performed during an endoscopy, permitted the complete aspiration of the fluid and the cytological analysis that showed numerous lymphocytes. The diagnosis of Intestinal lymphangiectasia was done, and the patient started the medium-chain triglycerides diet with significant improvement of her symptoms.

Conclusion

Intestinal lymphangiectasia is a cause of chronic diarrhoea that must be taken into account, even in old patients.

Introduction

Intestinal lymphangiectasia (IL) is a protein-losing enteropathy caused by congenital malformation or secondary obstruction of intestinal lymphatic vessels, which usually causes excessive protein loss in the intestine and malabsorption of both chylomicrons and fat-soluble vitamins. Thus patients present diarrhoea, oedema, hypoalbuminaemia, sometimes pleural effusion and ascites. The diagnosis is based on the upper gastrointestinal endoscopy showing diffuse scattered mucosal white blebs with typical histological finding of abnormal lymphatic dilatation. This article reports a case of IL with symptoms of chronic diarrhoea and an abdominal mass.

Case report

A 70-year-old woman came to our gastroenterological clinic for diarrhoea. This symptom began 8 months before; she referred increasing in frequency of evacuation (5–6 times a day), with watery stools mixed with mucus and sometimes with bright red blood. The evacuation was not painful, and neither abdominal pains nor urge faecal incontinence was referred. Furthermore she complained of retrosternal pain, which usually lasted a few minutes and receded spontaneously, without connection with eating. She was also suffering from itching in the superior limbs and neck for 20 days, without any visible skin lesion. Four months before, she had fever (body temperature 38°C–38.5°C) for 1 month associated with fatigue and lack of appetite, retreating with several and not specified antibiotic treatments. She was afflicted with gastroesophageal reflux, had hiatal hernia and treated fora Helicobacter pylori gastritis 20 years before; she took pantoprazole tablets. Furthermore she suffered from haemorrhoids. The patient had suffered from hypertension and chronic atrial fibrillation for 16 years, and she took diltiazemchloridate and calciparin. She had a cholecystectomy when she was 25 years old. Six years before, she underwent a right hip prosthes operation. She had family history of cholelitiasis and cardiovascular diseases. The patient smoked five cigarettes per day and drank about 26 g of alcohol (two drinks) per day. She was obese (body mass index: 34.5 kg/m²).

The clinical examination revealed tenderness in epigastrum and both iliac fossae, no palpable masses, no peripheral oedema. The complete blood count was normal except for a mild eosinophilia; the electrophoretic protidogram was also in normal range; no alterations of liver, pancreatic and thyroid function tests were found. Serum iron, calcium, glucose and electrolytes were within the normal ranges. The C-reactive protein was high (16.7 mg/L). Serum carcinoembryonic antigen (CEA) and carbohydrate antigen 19-9 (CA19-9) were in the normal ranges. Urinalysis, stool culture, research for Yersinia enterocolitica and stool ova and...
parasites exam were negative. Faecal calprotectin and antibodies for coeliac disease were normal, with IgA in the normal range. Ultrasound scan (US) showed a case of post-cholecystectomy with a mild dilatation of choledocus as vicariant gallbladder; the gastric wall was thickened, and a retropancreatic cystic mass, of about 2 cm, without vascular signals was found. Computed tomography (CT) scan reported minimal dilatation of intrahepatic bile ducts and common bile duct (7 mm). The pancreas was normal in size and shape; an exophytic and ovalar cystic mass of 3.4 cm, without sure continuity with the dorsal pancreas body, was found (Figure 1). This lesion did not show significant variations of enhancement in the different contrast phases. The radiologist supposed a cystic neoplasm of the pancreas and asked for a magnetic resonance imaging (MRI) scan to better characterise the mass. The patient refused the MRI scan, because she was afraid for her hip prosthesis and for the length of the examination. She then underwent an echoendoscopy (EUS) reporting that the ultrasound pancreatic aspect was compatible with fibro-adipose involution, and the Wirsung duct was regular without dilatation. On a level of pancreatic body, a gross complex cystic mass of 30 x 15 mm with internal septa was found (Figure 2). To better understand the diagnosis, a fine needle aspiration (FNA) of the liquid was performed for cytologic typing and CEA, CA19-9 and mucins test. The complete aspiration of the fluid was done as well as sample cells from the wall of the cyst, and about 6 cc of a citric yellow mucinous liquid was drained. The anatomopathological examination showed fibrin and numerous lymphocytes. The diagnosis of malabsorption due to IL was highly probable, so we started therapy with medium-chain triglyceride (MCT) diet, and we suggested that the patient eliminate fatty products, such as dairy products, butter and olive oil, and substitute those with a MCT oil. One month after MCT diet, the patient referred improvement of diarrhoea but still frequent retrosternal pain. She had so an oesophagogastroduodenoscopy that revealed no oesophageal lesions, a small hiatal hernia, bile pool and erosions in the stomach; the histological examination showed fragment of normal corpus and antrum gastric mucosa with mild congestion of lamina propria (Figure 3). The research for H. pylori was negative. At 2-month follow-up, the patient referred physical well-being and regular bowel movements, confirming the diagnosis.

**Discussion**

IL is an uncommon disorder and an important cause of protein-losing enteropathy caused by congenital malformation or obstruction of intestinal lymphatic drainage. All causes of elevated pressure of lymph drainage in the intestinal wall could lead to dilation and even rupture of the lymphatic vessels, which, in turn, results in the leakage of lymphatic fluid. As lymphatic fluid contains a lot of protein, fat and lymphocytes, leakage of lymph will cause hypoproteinaemia, lymphocytopaenia and decreased serum levels of immunoglobulin. IL was described for the first time half 50 years ago by Waldmann. In this case, the patient with oedema and hypoproteinaemia, with both hypoalbuminaemia and hypogammaglobulinaemia, were observed.

Depending on the cause of the disease, it can be classified into primary intestinal lymphangiectasia (PIL) or secondary intestinal lymphangiectasia, so the diagnosis of PIL must first exclude the possibility

---

**Figure 1:** Computed tomography (CT). Scan reported minimal dilatation of intrahepatic bile ducts and common bile duct (7 mm). The pancreas was normal in size and shape; an exophytic and ovalar cystic mass of 3.4 cm, without sure continuity with the dorsal pancreas body, is shown (red arrows).
Competing interests: none declared. Conflict of interests: none declared. All authors contributed to the conception, design, and preparation of the manuscript, as well as read and approved the final manuscript. All authors abide by the Association for Medical Ethics (AME) ethical rules of disclosure.


Figure 2: Echoendoscopy (EUS). On a level of pancreatic body, a gross complex cystic mass of 30 x 15 mm with internal septa is shown.

Figure 3: Oesophagogastroduodenoscopy (EGD). Cardias without mucosal alterations, no esophageal lesions, a small hiatal hernia. (B) Bile pool in the body of the stomach. Gastric folds flattened after-air insufflation, with normal thickness. (C) Antrum gastric mucosa with mild congestion and some erosions. (D) Duodenum with normal aspect.

Of secondary IL, e.g. other intestinal diseases (Crohn’s disease, Whipple’s disease, intestinal tuberculosis), lymphoma, sarcoidosis and scleroderma, radiation- and/or chemotherapy-induced retroperitoneal fibrosis or even a circulatory cause (e.g. constrictive pericarditis)⁴. Although PIL is more common in children, many cases were described in adults⁵,⁶. PIL affects equally boys and girls; the prevalence is unknown; it is generally diagnosed before 3 years of age. Patients with PIL often present bilateral lower limb oedema, sometimes with lymphedema, diarrhoea, malabsorption syndrome; pleural effusion and chylous ascites can also occur. If severe oedema is evident, facial and scrotal (or vaginal) involvement may occur⁴. As a consequence they usually show fatigue, abdominal pain, nausea, vomiting, weight loss, inability to gain weight, iron deficiency anaemia and obstructive ileus. Blood tests show often lymphopaenia, hypoalbuminaemia,
and hypogammaglobulinaemia due to lymph leakage from the ruptured lymph vessels. Patients can also develop hypocalcaemia secondary to failure to absorb fat and fat-soluble vitamins. Furthermore malabsorption of calcium, phosphate and vitamin D results in a high risk of osteoporosis and osteomalacia with decreases in bone mineral density and abnormal mineralisation of the protein matrix. Some cases of gastrointestinal bleeding were associated with IL, in one report massive bleeding caused by IL localised to the jejunum needed a surgical resection. Another study showed evidence of a strong association between small bowel angiodysplasia and lymphangiectasia, so this could be another explanation for intestinal bleeding and anaemia found in IL patients.

In this presented case, the first symptom referred was the diarrhoea with occasional bright red blood, but the patient suffered from haemorrhoid, so this is the most probable explanation for the presence of blood. Although diarrhoea is a symptom of malabsorption, no hypoalbuminaemia or other deficiencies were found in blood tests. The most important finding was the US imaging of a cystic retropancreatic mass. In literature, other cases of lymphangiectasia presenting as abdominal mass are reported, and in some of that cystic lymphangiomas were described.

Tests that evaluate proteinuria and rheumatic, neoplastic and parasitic infection can be used to exclude alternative diagnoses and the possibility of secondary IL.

IL typically affects the small bowel with an endoscopic appearance of white papules corresponding to the dilated lymphatics. The density of lymphangiectasia varies, and their size ranges from millimetre to centimetre. Histological examination of duodenum-jejenum and ileum biopsies usually establishes a definitive diagnosis and displays the presence of lacteal juice, dilation of mucosal lymphatic vessels without any evidence of inflammation and polyclonal normal plasma cells. Endoscopy may be negative when intestinal lesions are segmental or localised. In these cases, videocapsule endoscopy is a useful tool to detect the presence of the lesions, instead double-balloon endoscopy-assisted biopsy helps to make the histological diagnosis.

Other tests proposed to diagnose IL are albumin scintigraphy, which shows the albumin leakage into the bowel, CT scan showing diffuse, nodular, small bowel wall-thickening and oedema, which are a consequence of the dilated lymphatics or MRI scan.

These imaging scans are also important to exclude any other causes of elevated pressure of lymph drainage in the intestinal wall. We did not find any related diseases that could cause IL, after all several studies reported cases of duodenal lymphangiectasia not associated to other disorders and often without clinical evidence of malabsorption, in patients undergoing routine upper gastrointestinal endoscopy.

Low-fat diet associated with supplementary MCTs is the cornerstone of IL medical management. Because a partial block of the intestinal lymphatic system produces a loss of lymph into the lumen upon a high-fat diet, dietary fat restriction is considered to be the first choice of treatment, preventing the rupture of intestinal lymphatics and ensuing the protein and T-cell loss. The choice of MCT is justified because they are directly absorbed into the portal venous circulation and thus provide nutrient fat but avoid lacteal engorgement. We suggested the patient to reduce or eliminate dairy products and olive oil from her diet and to substitute it with an MCT oil to dress the food. Evidence showed that this kind of therapy, although not curative in all cases, improves the outcome and reduces the mortality in IL patients. This dietetic profile must be followed lifelong, because clinical and biochemical findings reappear after low-fat diet withdrawal. In patients not responding to a low-fat diet, enteral nutritional therapy (elemental, semi-elemental and polymeric diets) may be required. In a few very severe cases, total parenteral nutrition is warranted. When diet is insufficient to alleviate the symptoms, the use of octreotide, a synthetic analogue of the somatostatin hormone, has been found to be successful in improving protein-losing enteropathy, probably by speculative mechanisms including reduction in lymph flow and immunomodulatory action. Surgery may be successful when fibrotic changes of the small bowel cause partial mechanical bowel obstruction.

**Conclusion**

IL is a cause of chronic diarrhoea that must be taken into account, even in old patients. It is also important to think of this disease in the differential diagnosis of retropancreatic cystic mass, in order to avoid unhelpful surgical operation and emotional stress to patients. In the majority of cases, dietary treatment is simple and effective, giving a lifelong well-being.

**Consent**

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

**Abbreviations list**

CT, computed tomography; EGD, oesophagogastroduodenoscopy; EUS, echoendoscopy; IL, intestinal lymphangiectasia; MCT, medium-chain triglyceride; MRI, magnetic resonance imaging; US, ultrasound.

Licensee OA Publishing London 2013. Creative Commons Attribution License (CC-BY)

Chronic diarrhea and abdominal mass: A case of intestinal lymphangiectasia.

Case Report

References