

Case Report

Cystic Hygroma- Rare Cause of Protein Losing Enteropathy: A case report

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ABSTRACT

Protein-losing enteropathy is a rare complication of a variety of disorders characterized by excessive loss of protein into the gastrointestinal tract due to impaired integrity of the mucosa. Cystic hygroma is the cystic variety of congenital malformation of lymphatic system which is rare as a cause of protein-losing enteropathy (PLE). The clinical presentation of PLE is highly variable, depending on the underlying cause, but mainly consists of edema due to hypoproteinemia. We present a case of 8 year old girl presented with swelling in the right side of neck since birth and subsequently developed generalized oedema for the last one year. Her investigations showed hypoalbuminemia, pericardial effusion, ascites and cystic hygroma on lymph node biopsy and histopathology. Her upper GIT endoscopy revealed scattered white spots, having a snowflake like appearance in the mucosa of second part of duodenum, biopsy and histopathology showed diffusely dilated mucosal and sub-mucosal lymphatic vessels. Finally the case was diagnosed as a case of protein losing enteropathy due to Cystic hygroma.

Keywords: Cystic hygroma, Protein losing enteropathy, lymphangiectasia

1. INTRODUCTION:

Protein-losing enteropathy (PLE) is a rare condition characterized by loss of protein through the gastrointestinal tract, leading to reduced level of serum protein level. Incidence and prevalence are unknown. Hypoproteinaemia might be complicated by edema, ascites, pleural, and pericardial

effusions¹⁻⁴. Protein-losing enteropathies can be caused by a diverse group of disorders. The increase in intestinal leakage of plasma protein occurs via one of the two mechanisms- intestinal mucosal injury and increased lymphatic pressure. Mucosal injury with or without erosion/ulceration may be due to inflammatory bowel disease, celiac disease and many other causes. Increased lymphatic pressure in the gut may be due to granulomatous or neoplastic involvement of the lymphatic system or after dilatation of lymph vessels. Protein leak via the surface epithelium into the gut, as occurs in intestinal lymphangiectasia, congenital abnormalities of the lymphatic system, or in disorders of venous stasis such as congestive heart failure or constrictive pericarditis⁵. Intestinal lymphangiectasia (IL) is a rare protein-losing gastroenteropathy caused by malformation or obstruction of intestinal lymphatic drainage⁶. All factors causing elevated

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pressure of lymph drainage in the intestinal wall could lead to dilatation and even rupture of the lymphatic vessels which, in turn, result in the leakage of lymphatic fluid⁷. Cystic hygroma, a painless mass, can manifest anywhere in the body. Presentation is related to the complications or pressure effects of cystic hygroma. Due to its pressure effect in the lymphatic system that produces lymphatic obstruction, it may cause IL that ultimately may rupture and leakage of lymphatic fluid. As lymphatic fluid contains a lot of protein, fat and lymphocytes, leakage of lymph will cause hypoproteinemia, lymphocytopenia, and decreased serum level of immunoglobulin. Hypoproteinaemia may cause edema, ascites, pleural, and pericardial effusions⁸. The diagnosis of primary intestinal lymphangiectasia is based on the clinical manifestations and laboratory and pathologic findings. Diagnosis of IL is confirmed by the presence of intestinal lymphangiectasia based on endoscopic findings with the corresponding histology of intestinal biopsy specimens. On endoscopy, scattered white spots, which have been described as snowflake-like appearance, may overly the small intestinal mucosa⁹⁻¹⁰. Since the intestinal abnormalities often occur at the jejunoileum and the lesions may be segmental, dilated lymphatic could be not visible at endoscopy and biopsy may be negativ¹¹⁻¹³. Because of rarity of problem in children, we found it of academic interest to report the case of Protein losing enteropathy due to Cystic hygroma, who was admitted in Department of Paediatric Gastroenterology and Nutrition of Bangubandhu Sheikh Mujib and Medical University (BSMMU).

2. CASE REPORT

Sumaiya, an 8 years old girl, second issue of non consanguineous parents had right sided swelling of neck since birth and was diagnosed as a case of cystic hygroma at the age of one year on the basis of lymph node biopsy. She was admitted with the complains of generalized swelling of body and cough for the last one year. On examination she was ill looking, mildly pale, anicteric. She had a swelling over right side of neck and had bilateral pitting oedema (Figure 1). Her blood pressure was normal and bed side urine for albumin was nil. On systemic examination there were crepitations over both lung fields. She had ascites but no organomegaly. Her weight was 18 kg. Complete blood count showed neutrophilic leucocytosis with lymphopenia (WBC count 17×10^9 /cmm, neutrophil 84%, lymphocyte 5%). ESR was 20 mm in 1st hour. Her chest x-ray revealed nonspecific lung infection and bilateral minimal pleural effusion and echocardiography showed normal findings. Prothrombin time INR 1.08 (Normal 0.8 to 1.2), serum ALT 41 U/L, serum total protein 48 gm/l (normal 55-65 gm/l), and serum albumin 20 gm/l (normal 35-55 gm/l). Her Montoux test (MT) and antinuclear antibody (ANA) were negative and serum LDH was 488 U/l (Normal 120 to 225u/l). Renal function test including serum creatinine, serum electrolytes, 24 hours UTP and spot urinary protein creatinine ratio were within normal limit. Her fecal α 1-antitrypsin was 1.16 mg/gm (Normal <0.27 mg/gm). Upper GIT endoscopy was done which revealed scattered white spots,

having a snowflake like appearance in the mucosa of second part of duodenum. Biopsy was taken from affected mucosa and histopathology showed diffusely dilated mucosal and sub-mucosal lymphatic vessels. So with above background she was diagnosed as a case of protein losing enteropathy due to primary intestinal lymphangiectasia from cystic hygroma with respiratory tract infection. She was treated accordingly with general supportive measurers, parenteral antibiotics, albumin infusion and advised for medium chain triglyceride (MCT) based diet.

3. DISCUSSION

Protein losing enteropathy (PLE) is not a single disease but a complication of variety of disorders. Although enteropathy is defined as an intestinal disease, the term PLE is more liberally used including loss from the oesophagus and stomach as well¹⁴. Clinical manifestations of PLE are highly variable and are determined in part by the underlying cause. The main findings are reduced serum concentrations of albumin, gamma globulin, (IgA, IgG, IgM), fibrinogen, transferrin and ceruloplasmin. The hypoproteinaemia causes oedema, ascites, pleural effusion & pericardial effusions¹⁻⁴. This patient was initially diagnosed as a case of cystic hygroma. Subsequently the patient developed oedema, ascites, and respiratory tract infection. Laboratory examination showed neutrophilic leucocytosis, lymphopenia, hypoalbuminemia, bilateral non-specific lung infection on x-ray chest, upper GIT endoscopy revealed scattered white spots, having a snowflake like appearance in the mucosa of second part of duodenum, biopsy and histopathology showed diffusely dilated mucosal and sub-mucosal lymphatic vessels. In most patients the diagnosis of PLE depends on history, physical examination and clinical manifestations. However if necessary, PLE can be established by the detection of Alpha-1 antitrypsin in the stool sample. In this case fecal α 1-antitrypsin was also in favour of PLE¹⁵. However the detection of alpha-1 antitrypsin does not show the site of protein loss, furthermore it will not show a positive result when the stomach is the site of protein leakage¹⁸. Endoscopy can detect characteristic mucosal alteration like more prominent lymphatics with increased leakage of lymph into bowel lumen, seen as white spots on the mucosa which have been described as having a snow-flake like appearance¹⁷ which was obvious in this case.

Treatment of PLE consists of two components, maintenance of nutritional status & treatment of underlying cause. As there is no well-defined treatment guide line, a low long chain triglyceride (LCT) diet supplemented with medium chain triglyceride (MCT) and high protein is still the cornerstone of IL therapeutic management. MCT is a substitute for LCT are directly absorbed in the portal venous circulation and thus provide nutrient fat but avoid lacteal engorgement, thereby preventing their rupture and lymph loss. A recent study showed that 63% of the cases of IL implemented with diet control were sensitive to this treatment¹⁸. Albumin infusion given for transient correction of hypoalbuminaemia. It has been reported that two cases were successfully treated with

octreotide and endoscopic appearance of duodenum had turned nearly normal¹⁹. However there were also cases with no such improvement after octreotide therapy²⁰. The mechanism of octreotide in IL remains to be elucidated, and their efficacy in IL was variable and insufficiently evaluated.



Fig 1. Cystic hygroma in the neck.

4. FINANCIAL SUPPORT

Self

5. CONFLICT OF INTEREST

None

6. CONTRIBUTION BY AUTHORS

All authors contributed in the management of case.

W Mazumder and N Begum have written the case report and B Karim revised the manuscript.

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