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About A New Case Of Protein-Losing Enteropathy On Intestinal Lymphangiectasia

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Protein-losing enteropathy (PLE) is a syndrome where there is an excess loss of proteins in the gastrointestinal tract (GI). It can occur in many clinical conditions. Management of PLE is complex and challenging and requires a team approach. The characteristic finding in PLE is a loss of serum proteins into the gastrointestinal (GI) tract, resulting in hypoproteinemia with subsequent low oncotic pressure, leading to edema, pleural effusions, ascites, and malnutrition [1]. Erosive and non-erosive GI diseases, as well as vascular disorders that result in increased central venous pressure or mesenteric lymphatic obstruction, can lead to protein loss via the GI tract [1].

In this article, we describe a new case of PLE on intestinal lymphangiectasia revealed by ascitic-edematous syndrome.

I. Case Report

A 65-year-old woman with no medical history. The patient was hospitalized initially, one year ago, in pneumology for respiratory symptoms of productive cough and dyspnea associated with mild abdominal distension evolving for one year in a context of weight loss and apyrexia. Chest x-ray shows large right pleural effusion. Pleural puncture shows transudate liquid, Xpert TBK gene were negative, and cytology shows inflammatory and hemorrhagic nature, absence of cells suspicious of malignancy and pleural biopsy were non-specific fibroinflammatory remodeling and absence of granulomatous lesion and absence of signs of malignancy. The BK sputum was negative.

Then a digestive symptomatology was installed made of epigastric and right hypochondrium pain with important abdominal distension, hepatomegaly and oedemas of the lower limbs reaching to the ankles, taking up the bucket and the patient was anicteric.

Breast examination reveals orange peel appearance with asymmetrical breast volume hard and taut breasts with no palpable nodules. (Image 1)

Cervical Thoracic Abdominal CT indicates presence of multiple mediastinal adenopathies, an hepatomegaly, discreetly irregular contours, signs of right heart failure (dilated inferior vena cava +hepatic veins) with no signs of portal hypertension, 2 calcified granulomas and moderate peritoneal effusion.

Pelvic ultrasound shows no abnormalities apart from diffuse abdomino-pelvic fluid effusion.

Biologically a disturbance of the hepatic balance made of cytolysis and cholestasis and lymphopenia was detected.

Both viral serology (B and C) for hepatitis and autoantibodies were negative.

Liver biopsy reveals microscopic nodular regenerative hyperplasia of the liver with peliosis, sinusoidal dilatation and focal obliteration of portal veins, interlobular bile ducts are normally represented. This appearance suggests the need to look for underlying vascular pathology, in particular thrombosis of the hepatic veins, cardiac liver with absence of cirrhosis and dysplasia and immunohistochemical complement showing no vascular tumor proliferation and negative CMV antibody.

Echocardiogram was normal with preserved systolic function and pericardial effusion.

Mammary ultrasound shows an inflammatory mastitis, classified ACR BIRADS 3 and breast MRI shows mastitis classified as BIRADS 3 too with absence of suspicious contrast.

Hypoalbuminemia and hypoprotidemia were detected and 24H proteinuria were normal.

Alpha1 antitrypsin fecal clearance > 3600 (N<400), the diagnosis of PLE was confirmed.

About etiological assessment, esophagogastroduodenoscopy coloscopy were normal, and video capsule endoscopy showed jejunal lymphangiectasia (Image 2). We couldn't make biopsy because of the distal and patchy localization of lymphangiectasia to the jejunum and ileum.

The patient was put on high-protein diet 2-3g/kg/day and low-fat diet and thromboprophylaxis.

Two months later, the patient was well with reduced oedema, and her weight had decreased. Protein levels and liver function improved.

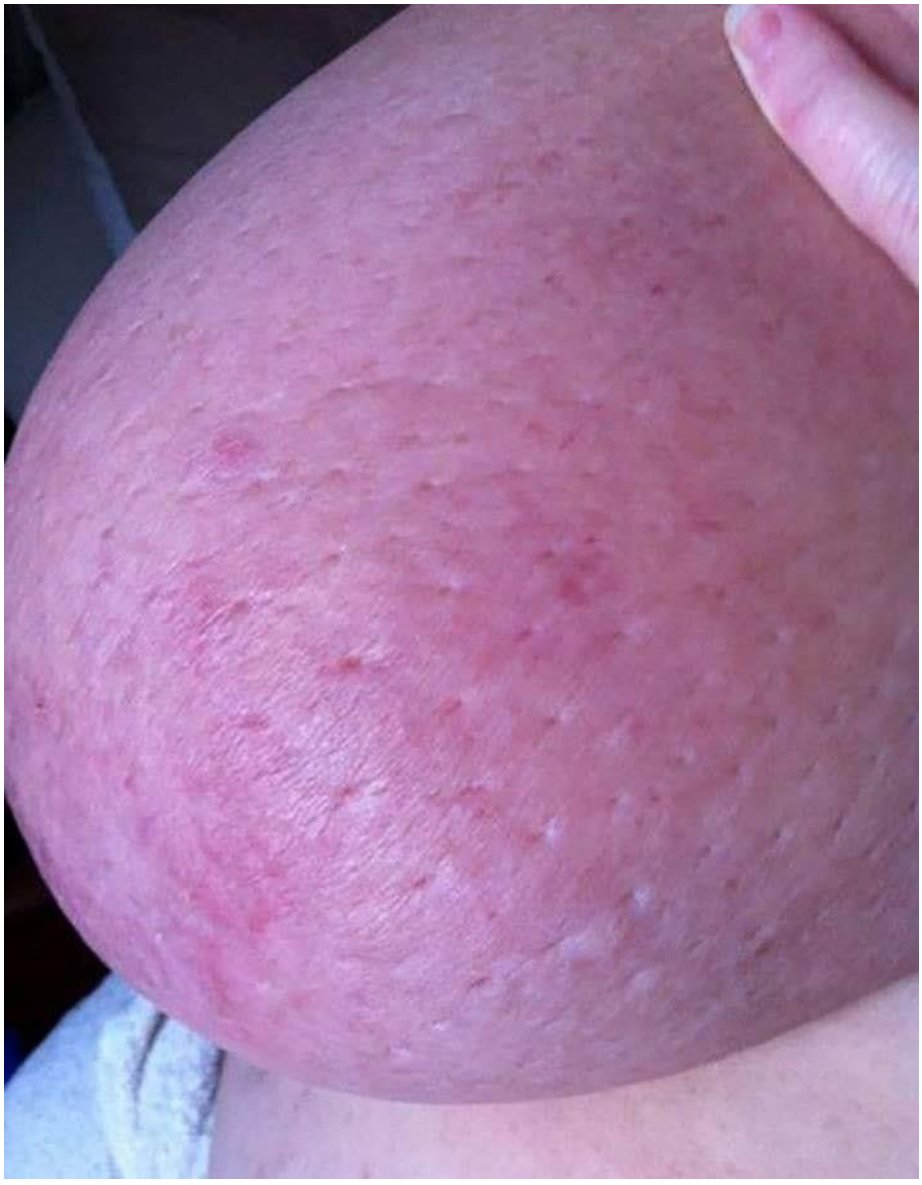

Image 1 : Orange Peel Appearance Breast

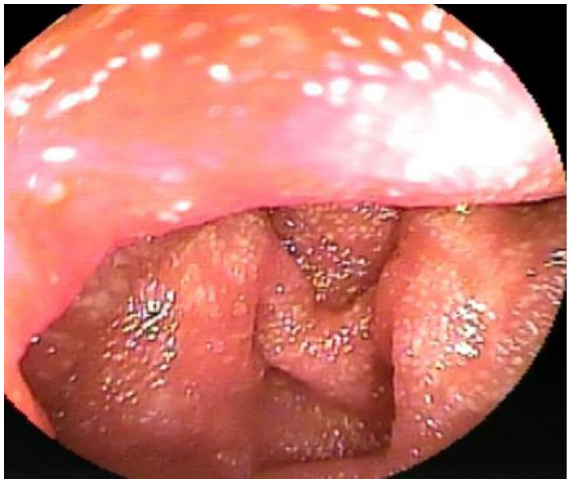

Image 2 : Video Capsule Endoscopy With Appearance Of Intestinal Lymphangiectasia

II. Discussion

PLE is not a well-defined clinical entity, but rather a manifestation of a wide variety of gastrointestinal and extra-intestinal diseases. Three main mechanisms are involved, allowing a pragmatic etiological classification: lymphatic leakage from intestinal lymphangiectasias; exudation from digestive ulcerations; and disruption of the epithelial barrier without impediment to lymphatic drainage or patent ulcerations. [2].

The three main groups of disorders that cause excess protein loss in stools are :

Primary Erosive/Ulcerative gastrointestinal Disorders :

This group of conditions includes inflammatory bowel diseases (both ulcerative colitis and Crohn disease), gastrointestinal malignancies, any erosions or ulcers of stomach or duodenum, *Clostridium difficile* colitis, carcinoid syndrome, graft vs. host disease.

Non-Erosive/Non-Ulcerative Gastrointestinal Disorders

This group includes topical Sprue, celiac disease, Menetrier disease, amyloidosis, cutaneous burns, eosinophilic gastroenteritis, bacterial overgrowth, Intestinal parasitic infections, Whipple disease, collagenous colitis, AIDS, mixed connective tissue diseases, systemic lupus erythematosus (SLE), rheumatoid arthritis (RA).[3][4]

Disorders Causing Increased Interstitial Pressure or Lymphatic Obstruction :

This grouping can be due to primary intestinal lymphangiectasia, right-sided heart failure, constrictive pericarditis, congenital heart disease, Fontan procedure for single ventricle, cirrhosis with portal hypertension gastropathy, hepatic venous outflow obstruction, mesenteric tuberculosis or sarcoidosis, retroperitoneal fibrosis, lymphoenteric fistula, lymphoma, and thoracic duct obstruction.[5][6][7]

Diagnosis of protein-losing enteropathy should be suspected in patients with hypoproteinemia once the other common causes like severe malnutrition, nephrotic syndrome, or chronic liver diseases have been ruled out. Since the protein loss in PLE occurs independent of their molecular weight, these patients have low albumin and low globulins in their serum. If there are isolated low serum albumin and normal serum globulins, then alternative causes should be considered.

Clinical examination and digestive endoscopic investigations are central to the orientation and etiological diagnosis.

Once the diagnosis of PLE is established with increased A1AT clearance, further workup is needed to find the underlying etiology and to guide treatment. Important consideration should be given to the history and physical examination of the patient. Detailed history helps in narrowing down the list of conditions causing hypoproteinemia and PLE and guides the approach towards further work up. Basic labs like complete blood count with differential, liver function tests, renal function tests should be performed in all patients. In cases where symptoms suggest gastrointestinal causes of protein loss, tests for celiac disease, infectious work for chronic intestinal infections, appropriate imaging of abdomen and pelvis, upper and lower gastrointestinal endoscopies with biopsies, capsule endoscopies (if upper and lower endoscopies are unremarkable) should be performed. Autoimmune workup should be ordered if systemic lupus erythematosus (SLE) or rheumatoid arthritis (RA) is suspected. Echocardiogram and workup on the lines of heart failure should be done if cardiac causes of PLE is suspected. Fontan procedure in patients with congenital heart disease is a well-known cause of PLE and has been extensively studied. [8]

Treating the underlying pathology is the mainstay of treatment. Besides this, dietary modifications also play a critical role in the management of protein-losing enteropathy. Diet rich in protein and medium chain triglycerides and low in fat is considered the best diet in this condition. Patients may require 2 to 3g/kg/day of protein. Replacement of micronutrients, electrolytes, and vitamin deficiencies should occur as appropriate.

If heart failure is the cause of PLE, then optimization of heart failure medications needs to be done. Diuretics are an option for symptoms of anasarca and fluid overload. Pericardiectomy can help in constrictive pericarditis. Treatment with immunosuppressive medications should be the approach in inflammatory bowel disease, SLE, RA, and other inflammatory conditions. Treat parasitic infections if they are the cause of PLE. Surgical resection can be a consideration in Menetrier disease[9] and refractory inflammatory bowel disease. Octreotide has been considered beneficial in primary intestinal lymphangiectasia and Menetrier disease by decreasing lymphatic pressure and reducing intestinal protein loss. Budesonide and corticosteroids may be helpful in eosinophilic gastroenteritis and some cases of PLE due to Fontan procedure.[10]

Routine monitoring is advisable after initiation of treatment in the form of checking micronutrients and vitamin deficiencies, serum protein levels, and A1AT clearance.

III. Conclusion

PLE is a clinicobiological entity whose etiology may be digestive or extra-digestive. Positive diagnosis is based on elevated alpha-1 antitrypsin (A1AT) clearance.

Dietary measures are always indicated in conjunction with treatment of the causal mechanism.

Prognosis is variable and dependent on the underlying cause of protein-losing enteropathy. If there is a successful treatment of the underlying cause, then it can lead to complete resolution of PLE.

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