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Case Report

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PRIMARY INTESTINAL LYMPHANGIECTASIA (WALDMAN'S DISEASE): A CASE REPORT

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INTRODUCTION

Primary intestinal lymphangiectasia or Waldmann's disease is due to lymphatic dilatation with leakage of lymph into the intestinal lumen responsible for hypoprotidemia, chronic diarrhea due to exudative enteropathy, and edema.It is a disease usually revealing itself in childhood, sometimes with growth retardation in severe forms and of which there are some familial forms. While edema is common in Waldmann's disease and is related to hypoproteinemia, lymphedema is less common but can affect the lower and upper limbs(1). In the course of the disease, they usually occur after the digestive involvement which is symptomatic and in the foreground. We report a case of lymphoedema of the limbs evolving for several years and considered as primary, before the diagnosis of paucisymptomatic Waldmann disease was made.

KEYWORDS: Waldmann's disease, primary intestinal lymphangiectasia, hypoalbuminemia.

OBSERVATION

This is a 26-year-old patient, followed since childhood for hypo-albuminemia with intermittent liquid diarrhea, hospitalized twice at the age of 2 months and 9 months for an oedemato-ascitic syndrome, where she had benefited from albumin infusions, she was put on diuretics with a good evolution and then the patient was lost from sight.

She was operated at the age of 24 years for a hydropneumothorax with good clinical and biological improvement.

In front of the reappearance of liquid diarrhoea, the patient had consulted in our training. The general examination found a stable patient on the HD and respiratory plan, apyretic with bilateral OMI not taking the cup the abdominal examination found a flexible abdomen breathes normally, no sensitivity, no dullness, a scar under umbilical. The pleuropulmonary and cardiovascular examinations were unremarkable.

A biological check-up was requested which showed a normocytic normochromic anemia with a hemoglobin of 10.4 g/dl VGM 86. 4 fL CCMH at 32 fL, correct leukocytes at 5880/uL PNN at 3234/uL lymphocytes at 1830/uL a correct platelet count at 353000 as part of the etiological work-up: A lipid work-up was requested which showed a triglyceride level : 0.38 g/L, Total cholesterol 1; 31g/L, HDL cholesterol 0,35 g/L, LDL cholesterol 0,88 g/L, a correct Calcemia at 88, a magnesia at 15, a correct albuminemia at 33g/L, a LDH level at 157 IU/L, beta-2-microglobulin at 2,82mg/L, an alpha-1-antitrypsin dosage which came back 1,48g/L with an increase in clearance of alpha-1-antitrypsin higher than 1800 ug/g, the search for blood in the stools was negative.

Still within the framework of the etiological assessment, an endoscopic assessment was carried out: an eosogastro-duodenal fibroscopy (Figure 1): which showed a very obvious duodenal lymphangiectasia, and an ileo-coloscopy (Figure 2): which showed a very obvious ileal lymphangiectasia, the anatomo-pathological examination found a chronic non-specific duodenitis, normal fundic mucous membrane and an edematous colitis.

An abdominal and pelvic CT scan was performed, which did not show dilatation of the bowels or thickening of the mucosal folds.

A thoracic MRI was requested as part of the etiological work-up, which did not reveal any ectasia of the thoracic duct, with the presence of a small bilateral pleural effusion on the right.

The association of lymphoedema of the limbs, visceral

effusions, exudative enteropathy and the preceding biological abnormalities suggested the diagnosis of primary intestinal lymphangiectasis or Waldmann disease.

The clinical course was marked by improvement of lower limb oedema and diarrhoea after the introduction of a well-established diet low in heavy-chain triglycerides and very regular follow-up with a dietician. The association of lymphoedema of the limbs, visceral effusions, exudative enteropathy and the preceding biological abnormalities evoked the diagnosis of primary intestinal lymphangiectasias or Waldmann disease.

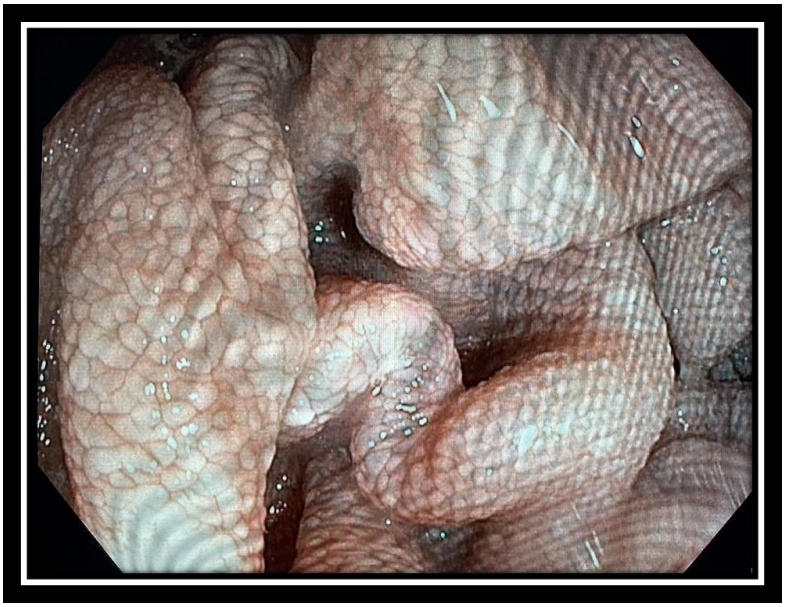

Figure 1: Fibro-oesogastro-duodenal showing multiple milky lesions corresponding to lymphangiectasias.[2]

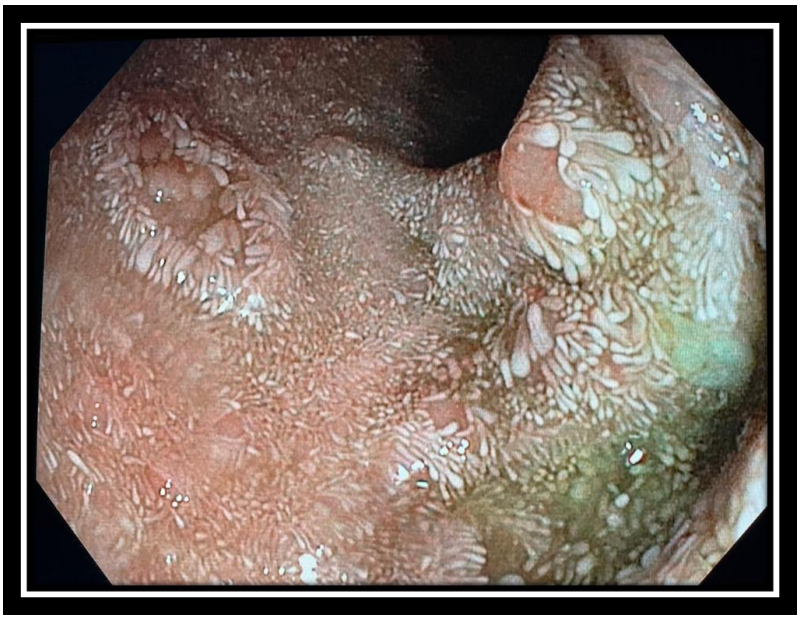

Figure 2: Ileo-colonoscopy showing a very obvious lymphangiectasia at the ileal level.[2]

DISCUSSION

The initial description of primary intestinal lymphangiectasias was made by Waldmann in 1961 and included twelve patients with hypo-albuminemia, hypo-gammaglobulinemia, edema of the lower limbs appearing before the age of 30 years, digestive disorders such as diarrhea and, in four cases, pleural and peritoneal chylous effusion.[3] A few years later, the same author completed the picture by associating lymphopenia, a lower reactivity to skin antigenic tests and the absence of rejection of heterologous skin grafts.[4]

The pathogenesis of Waldmann disease remains unknown. The primary character of intestinal lymphangiectasias can only be affirmed after eliminating secondary causes of digestive lymph loss: malformations of the lymphatic ducts, diseases of the lymph nodes

(lymphoma, metastases, tuberculosis, filariasis, Whipple's disease), compression of the lymphatic ducts (tumors, retroperitoneal fibrosis, mediastinitis), cardiopathies with venous hyperpressure, thrombosis of the inferior vena cava, lympho-intestinal fistulas, iatrogenic lesions of the lymphatic system (lymph node excision, radiotherapy). The disease most often begins in childhood but can be congenital or in adolescence. It may be suspected on prenatal ultrasound by the presence of fetal edema.[5] Conversely, a revelation of the disease in adulthood is also possible. Both sexes are affected in a comparable manner.

Edema is constant and often asymmetrical, whereas chylous effusions are rare. These edemas, localized to the lower limbs, may also affect the upper limbs. Mostly related to hypoalbuminemia, they are diffuse, white and soft. However, true lymphedema may be associated with Waldmann disease as well as other lymphatic abnormalities (cystic lymphangioma, splenic lymphangiectasia).[5,6] These various abnormalities are consistent with diffuse lymphatic disease. Lymphedema may be present at the time of diagnosis of primary intestinal lymphangiectasias or later in the course. The presence of lymphedema preceding the diagnosis of Waldmann disease by several years, as in our observation, is very unusual. Lymphoscintigraphy with technetium-99 colloid injection, which has replaced direct lymphography, may show various diffuse abnormalities of the lymphatic system: hypoplasia of the lymphatic vessels of the limbs, agenesis or hypoplasia of the lymph node relays, subdermal lymphatic reflux in the territories affected by lymphedema.[4]

The digestive disorders are most often summarized by intermittent moderate diarrhea, the origin of which is exudative enteropathy. The severity of the disease is probably due to the extension of the lymphangiectasias on the digestive tract. This is confirmed by the increase in α1-antitrypsin clearance and the normality of the D-xylose test. Steatorrhea is inconstant. Duodenal biopsies confirm the diagnosis by finding lymphangiectasias in the duodenojejunal mucosa and submucosa, the rupture of which would be responsible for the lymphatic leak. Sometimes, the macroscopic aspect can be evocative with a whitish and puffy aspect of the digestive villi. Multiple biopsies are required because the lesions may be very localized. There is no villous atrophy (2.7) or inflammatory infiltrate. Other morphological examinations are of little value. Thoracoabdomino-pelvic CT may show dilatation of the small intestines and a thickened mucosal fold.

The leakage of lymph through the digestive tract is responsible for hypoalbuminemia, hypogammaglobulinemia, hypocalcemia and lymphopenia.

Treatment is symptomatic. In case of disabling diarrhea, a diet low in long-chain fatty acids, enriched with medium-chain fatty acids that pass directly into the bloodstream, and fat-soluble vitamins A and E are prescribed. Serous effusions are evacuated and pleural talcage may be performed in case of frequent recurrences. The treatment generally allows the regression of biological abnormalities, but in some cases biological stigmata of lymphatic leakage may persist, whereas the clinical signs may disappear if the diet is strict. Moreover, normalization of lymphocyte count does not always restore normal immune function. Treatment with subcutaneous octreotide has also been reported in a few cases where clinical signs persisted on diet. Lymphedema is not sensitive to the fat-free diet but should be treated with the usual compression techniques.

CONCLUSION

Waldmann's disease is a chronic disease requiring a prolonged and restrictive strict hypolipid diet, supplemented with medium-chain triglycerides and fat-soluble vitamins. Quality of life is often impaired by the presence of edema and/or lymphedema, and asthenia is frequent. The evolution can be complicated by episodes of serous effusions (pleural, pericardial) with a more or less important functional repercussion and which can in the most serious forms involve a vital risk. The occurrence of non-Hodgkin's lymphoma is more rare. Prolonged clinical and biological monitoring is necessary

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