Intestinal involvement in Waldenström’s Macroglobulinemia

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Abstract

We describe the case of a patient with Waldenström’s macroglobulinemia in which routine ultrasound examination revealed diffuse thickening of the bowel walls concomitant with a hyperemic circulation, mesenteric infiltration, intraabdominal lymph nodes enlargement, ascites and signs of portal hypertension; these findings were in discrepancy with the absence of a significant clinical pattern. This case underlines the importance of the ultrasound scan in Waldenström’s macroglobulinemia, even in the presence of a paucisymptomatic patient.

Key-words: Waldenström’s macroglobulinemia, intestinal involvement, lymph nodes, ascites, portal hypertension

Rezumat

Se prezintă cazul unei paciente cunoscute cu macroglobulinemie Waldenström. Examinarea ecografică de rutină a depistat ingroșarea difuză a periților intestinali cu hiperemic, infiltrarea mezenterului, adenopatii intraabdominale, ascită și semne de hipertensiune portală în discordanță cu tabelul clinic paucisymptomatic. În concluzie, se subliniază necesitatea ca examenul ecografic să facă parte din examinarea de rutină a pacienților cu boală Waldenström, chiar în absența unei simptomatologii specifice.

Key-words: macroglobulinemia Waldenström’s, afectare intestinală, adenopatie, ascită, hipertensiune portală

Introduction

Waldenström’s macroglobulinemia (WM) is a B cells lymphoproliferative disorder characterized by bone marrow infiltration with lymphoplasmocytes and by the presence of monoclonal paraproteinemia with IgM [1]. The clinical pattern is variable, the signs and symptoms being, usually, the consequence of bone marrow infiltration and blood hyperviscosity. Intestinal involvement in WM is rare, caused by deposition of macroglobulinemic paraproteinemia at the level of intestinal microvilli, both interstitial and in lymphatic capillaries with their secondary obstruction. As a consequence, there is an intestinal mucosal thickening with a severe malabsorption syndrome [2]. Apart from some older autopsy reports regarding intestinal involvement in MV [3], endoscopic aspects of intestinal involvement in WM have been noted at the level of the duodenum and terminal ileum [4]. In a recent communication the whole endoscopic aspect of intestinal involvement observed during endoscopic capsule examination was described [5]. However, this aspect is not pathognomonic. A similar pattern can be found in other medical conditions such as primary lymphangiectasia, Whipple’s disease or in any situations associated with severe lymphatic obstruction. Additional tests are required for differential diagnosis: intestinal biopsy and immunofluorescence examination.

Case presentation

We present the case of a 62 year old female patient diagnosed with Waldenström’s disease from 2005, who was
admitted for clinico-biological reevaluation. At the moment of diagnosis the biological data revealed anemia, an inflammatory syndrome with elevated ESR, positive Coombs test, modified aspect of immuno-electrophoresis (raised monoclonal IgM component), altered bone marrow biopsy (hypercellular marrow due to the presence of multiple lymphoid infiltrates). Family history was irrelevant. The patient was a nonsmoker and her consumption of alcohol was low. From the diagnosis until the present time she has been treated with chemotherapy. The physical examination revealed a good nutrition, normal temperature, mild mucosal and skin pallor, bilateral small lymph nodes enlargement in latero-cervical areas and bilateral large adenopathy in inginal regions having a diameter of 2-3 cm (unpainful, mobile and dense lymph nodes). The abdomen was enlarged with dullness at percussion in declining areas. The palpation revealed hepatomegaly with inferior margin at 2-3 cm under costal ribs, splenomegaly with inferior edge at 2 cm under costal ribs. There was no changes regarding the respiratory system, cardiovascular system or central and peripheric nervous system.

The laboratory tests revealed a mild normocromic normocytic anemia (Hb = 8.5 g/dl, Ht = 26.2%), the presence of an inflammatory syndrome (ESR = 64 mm/h), hyperuricemia (9.5 mg/dL), hyposideremia (35 µg/dL), hyperbilirubinemia (total bilirubin = 1.73 mg/dL, direct fraction = 0.48 g/dl) and significant hypergammaglobulinemia (total protein = 9.87 g/dL, albumine = 51.04%, gammaglobulins = 42.25%).

Abdominal ultrasound examination identified a dilated portal vein (16 mm) (fig. 1), splenomegaly (fig. 2), moderate ascites, small retroperitoneal (periaortitic) adenopathy, multiple mesenteric adenopathy (fig. 3), images of bowel infiltration (fig. 4-8), especially on the left side of the abdomen, hyperemic circulation in the bowel wall (fig. 9); the liver, gallbladder and biliary tree appeared normal.

![Fig. 1. Dilated portal vein (16.1 mm) representing portal hypertension; normal liver ecostucture.](image1)

![Fig. 2. Splenomegaly.](image2)

![Fig. 3. Color doppler examination for differential diagnosis between intraabdominal lymph nodes and mesenteric vessels.](image3)

**Discussions**

The gastrointestinal complications in Waldenström’s macroglobulinemia are rare [6, 7]. In the case presented above, there were no signs of intestinal malabsorption despite the thick bowel wall identified during ecography. The only manifestation eventually related to this infiltration of the bowel wall was hyposideremia and its related anemia. In this case we did not perform an upper gastrointestinal endoscopy or colonoscopy with biopsy. The studies published in literature showed that patients with a similar clinical situation have had a lymphoplasmocytic infiltration of the intraabdominal lymphatic system (lymph nodes and lymphatic vessels, including those in the intestinal wall) together with interstitial infiltration of the intestinal microvilli [2, 8]. However, those patients had clinical signs of malabsorption: diarrhea, steatorrhea, protein losing enteropathy [9]; histologically, they also had
In our particular case, the patient had no signs of malabsorption, but because the disease was not at the initial period, but in a stable, treated phase.

Regarding the presence of portal hypertension syndrome, this was also described in the literature, although many years before, being in fact a presinusoidal hypere regeneration of the liver, that in such a situation there is a liver infiltration of the mesenteric lymphatic and interstitial deposits of immunoglobulin M. Infiltration of lymph nodes involvement as a consequence of hyperplasia due to the abnormal blood viscosity. Furthermore, lymphangiectasia in Waldenstrom’s macroglobulinemia may also be because the disease was not at the initial period, but in a stable, treated phase.

Fig. 4, 5, 6, 7, 8. Bowel loops with hypoechoic concentric thick wall, surrounded by hyperechogenic mass (infiltrated mesentery).

Fig. 9. Power doppler examination: bowel wall hyperemia and perimesenteric adenopathy; left renal cyst.
portal tracts with lymphoplasmocytoid cells of the same type as those in the bone marrow. The mechanism of portal hypertension is presumed to be double: (1) an increased of blood resistance at the infiltrated portal tracts, and (2) an increased of blood inflow from the splenic vein due to the presence of splenomegaly. Portal hypertension was, also, described in relation to other myeloproliferative disorders or lymphoma [12-14].

The presence of ascites in Waldenström’s macroglobulinemia is extremely rare, until now being described only in three cases: one case of WM with pleurisy and ascites, another case in which a malignant ascites occurred during WM transformation in immunoblastic lymphoma, and one last case of WM with the presence of ascites from the onset of the disease [15-17]. The mechanism of ascites is related to the presence of portal hypertension, caused by presinusoidal obstruction due to the blood hyperviscosity. The clinical signs of blood hyperviscosity are epistaxis, confusion, dizziness, congestive heart failure; ascites is another, very rare, sign in this clinical syndrome.

Conclusions

Ultrasound examination of the abdomen requires to be performed regularly in patients with Waldenström’s disease, even in the absence of a specific symptomatology, because there can be signs of intestinal and extraintestinal involvement of an infiltrative pattern, together with lymph node enlargement and signs of portal hypertension.

References