Letter to the Editor

Waldenström macroglobulinemia and portal hypertension

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TO THE EDITOR

Sir, Waldenstrφm macroglobulinemia is a clinicopathologic symptom in which a B-cell neoplasm involving the bone marrow, usually lymphoplasmacytic lymphoma, is associated with immunoglobulin M paraprotein in serum. Waldenstrφm macroglobulinemia may infrequently involve liver and in the majority of cases has been related to a subsequent diagnosis of hepatocellular carcinoma. We would like to report herein a successful management of a patient with symptomatic portal hypertension related to Waldenström macroglobulinemia.

A 56-year-old female patient diagnosed with Waldenstrom's macroglobulinemia five years ago, was admitted to our Department due to upper gastrointestinal (GI) tract bleeding. Patient medical history was unremarkable except of fludarabine for Waldenstrom macroglobulinemia. During hospitalization an upper GI endoscopy was performed revealing third degree esophageal varices and congestive gastropathy, findings suggestive of portal hypertension. Esophageal banding was successfully performed in esophageal varices. At that time patient was switched chemotherapy to an anti-CD20 monoclonal antibody. Routine investigation for other possible underlying diseases related to portal hypertension was negative and spleen was in upper normal limits. A liver biopsy was performed showing mild hepatitis with remarkable lymphocyte and plasma cell infiltrations and stage 3 fibrosis ac-

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cording to Ishaak grading system. Patient was discharged in a stable condition and was started on endoscopic follow up for the esophageal varices while the administration of antiCD20 monoclonal antibody was continued. On the two-year follow up patient remains in hematological stability and with a remarkable compensation of portal hypertension without further recurrence of esophageal varices bleeding.

This is an infrequent patient case of portal hypertension related to Waldenstrφm macroglobulinemia, which was successfully treated with esophageal varices banding and anti-CD20 monoclonal antibody. On current follow up portal hypertension portal hypertension is under complete compensation.

Except of liver involvement in the form of hepatocellular carcinoma, Waldenstrφm macroglobulinemia may be infrequently related to portal hypertension. In fact, only two cases with portal hypertension related to Waldenstrφm macroglobulinemia have so far been described.⁷⁻⁸ Lymphadenopathy in Waldenstrφm's macroglobulinemia causing extrahepatic portal hypertension was the underlying mechanism in one case while co-existing liver cirrhosis related either to hepatitis C⁹ or alcoholic liver disease¹⁰ were the triggering factors in other cases. It is also noteworthy that chronic liver disease may manifest as Waldenström's macroglobulinemia.¹¹

To conclude we reported a rare case of variceal bleeding due to portal hypertension related to Waldenstrφm macroglobulinemia, which was successfully treated with anti-CD20 monoclonal antibody and endoscopic banding.

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