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Chronic lymphocytic leukemia with massive ascites: An unusual presenting manifestation

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Dear Editor,

Chronic lymphocytic leukemia (CLL) is a lymphoproliferative disorder that usually manifests as lymphocytosis, with or without lymphadenopathy, organomegaly, anemia, thrombocytopenia due to marrow infiltration, and autoimmune phenomena. Ascites is rare in CLL patients at the time of diagnosis. Development of ascites in CLL has been described at the time of relapse or transformation into prolymphocytic leukemia or Richter's syndrome. Here, we report a case of CLL, who presented with massive ascites at presentation.

A 52-year-old female presented with a history of fatigue, weight loss of 4 months' duration, and abdominal swelling of 4 weeks' duration. On examination she had massive ascites and splenomegaly, while there were no clinical signs of chronic liver failure. Systemic examination was unremarkable; also, there was no peripheral lymphadenopathy. Ultrasonography of the abdomen showed gross ascites, splenomegaly (10 cm), normal liver size and echotexture, and no lymph node enlargement. A computed tomography scan of the abdomen showed splenomegaly and ascites. Her lab investigations revealed a hemoglobin of 12.5 g/dL, total leukocyte count of 90 × 109/L, and a platelet count of 180 × 109/L. Peripheral blood smear examination showed 90% lymphoid cells, which were CD5+, CD23+, CD19+, CD20+, CD45+, FMC7-, and CD3- on flow cytometry suggestive of CLL. Renal and liver function tests were within normal limits. Mantoux test was negative. Abdominal paracentesis yielded fluid showing white blood cell count of 840/dL, with 10% polymorphs and 90% lymphocytes. Morphological and immunophenotypic analysis of ascitic fluid was consistent with CLL. Gram stain, culture, and stain for acid-fast bacilli in the ascitic fluid was negative. Serological tests for hepatitis B, hepatitis C, and human immunodeficiency virus were negative. An upper and lower gastrointestinal endoscopy was normal. A diagnosis of CLL, Rai Stage II was made and the patient was started on 3 weekly CVP regimen, consisting of cyclophosphamide 1 g intravenous (IV) bolus (D1), vincristine 2 mg IV push (D1), and prednisolone 100 mg for 5 days. She had a significant reduction in ascites and improvement in general well-being within 4 weeks of starting treatment and no requirement of therapeutic paracentesis after four cycles. At present, she has completed eight cycles of CVP and her disease is in complete remission.

Ascites is a common manifestation of some advanced malignancies arising from the gastrointestinal tract, breast, and ovary. However, it is an unusual feature in chronic lymphoproliferative disorders and has been seen only in a few cases of CLL during Richter's transformation.[1,2] Ascites as an initial manifestation of CLL has been rarely reported.[3,4] Davis *et al*., reported chylous ascites in a patient

of CLL and Siddiqui *et al*., reported ascites in a 75-year-old male, both of whom were treated with the CVP regimen.[3,4] The pathogenesis of ascites in CLL is not known; however, it is hypothesized that abdominal lymph nodes and/or peritoneal deposits in CLL give rise to ascites, similar to what is seen in patients with non-Hodgkin lymphoma. The two cases described in the literature were both associated with abdominal lymphadenopathy and/or peritoneal deposits, unlike our case, where there was no evidence of the same; this suggests that there may be other mechanisms responsible for ascites in CLL. It has been demonstrated that ascitic fluid contains increased levels of vascular endothelial growth factor and vascular permeability factor produced by lymphoma cells which stimulate vascular leak or portal system lymphatic obstruction due to extensive lymphatic infiltration, leading to development of symptomatic ascites, and this might be the reason in our case.[5] These findings may also be seen with liver disease and portal hypertension, ovarian carcinoma, gastric cancer, colon cancer, and breast cancer. However, liver cirrhosis and portal hypertension, and other malignancies were ruled out with required investigations in our case.

The presence of ascites at baseline in a case of chronic lymphoproliferative disorder might sometimes mislead the physician into considering other more common conditions. This case highlights the fact that ascites can be a rare presenting feature of CLL.

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