

Ascites as a Manifestation of Hairy Cell Leukaemia Variant

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DOI: [10.36347/SJMCR.2019.v07i11.010](https://doi.org/10.36347/SJMCR.2019.v07i11.010)

| Received: 09.11.2019 | Accepted: 17.11.2019 | Published: 24.11.2019

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Abstract

Case Report

Hairy cell leukaemia is a blood neoplasm that represents approximately 2% of all lymphoid leukaemias. When present, symptoms include asthenia, abdominal pain, and unintended weight loss. We present the case of a 90-year-old male patient with a history of hairy cell leukaemia variant that went to the Emergency Room due to an increase in abdominal size and swelling of the lower limbs. He was admitted to the Internal Medicine Department for study and compensation of ascites condition. Undeniably, the most common cause for ascites is portal hypertension related to liver cirrhosis. However, there are malignant and/or infectious causes that should always be ruled out. In the case presented, after ruling out the most common causes for ascites, it was determined that the ascites was secondary to hairy cell leukaemia, despite generally presenting an indolent course.

Keywords: Ascites; hairy cell leukaemia variant.

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INTRODUCTION

Hairy cell leukaemia predominates in Caucasian males (4:1), with the average age of presentation being 52 years; it almost never affects children. Presentation is usually indolent, particularly in the early stages and can only be diagnosed through routine blood tests. When symptoms are present, they include:

- Asthenia;
- Abdominal pain (commonly due to splenomegaly);
- Unintended weight loss [1].

Diagnosis requires a bone marrow exam that identifies hairy cells. Besides the classic form, there is the variant form, which represents 10% of the patients with hairy cell leukaemia and whose prognosis is worse since it responds less effectively to the current treatments [1, 2]

Hairy cell leukaemia variant, in particular, is a rare B-cell lymphoproliferative chronic disorder with morphological features intermediate between hairy cells and prolymphocytes [2, 3]

Definitive diagnosis of both is done through immunophenotyping; both express CD20, CD22, CD11c, and CD103; however, contrary to hairy cell leukaemia that expresses CD123 and CD25, hairy cell leukaemia variant lacks expression of CD25 and is normally negative for CD123[4].

CASE REPORT

The patient is a 90-year-old, totally autonomous, male that went to the Emergency Room due to an increase in abdominal size and swelling of the lower limbs that had been evolving for around 10 days. He admitted dyspnoea and asthenia that was gradually worsening. His relevant personal history included hairy cell leukaemia variant diagnosed in February 2014, after having been admitted for an etiological study of splenomegaly. He had follow-up outpatient Hematology consultations every 4 months. After the diagnosis, he was proposed for a splenectomy, which did not take place due to his advanced age, co-morbidities and the observed clinical stability. The option made was for vaccination and close echocardiographic and analytical control.

During the objective exam, he was aware and cooperative. Apyretic, cyanotic and anicteric. Pink and hydrated mucous membranes. Normal heart rate. Eupneic in ambient air and cardiopulmonary auscultation showing no relevant changes. Swollen, slightly depressible, abdomen in moderate tension. He also showed moderate sensitivity to palpation, although without defence. Swollen lower limbs with godet +++/+++.

The chest X-ray revealed hilar enlargement, an increase in the cardio-thoracic (CTI) index, although no condensation or apparent effusion.

Analytically without leukocytosis or neutrophilia, with a Hg of 12g/dL and a platelet count of 111000. No changes in the clotting study. Kidney function with 2.18 mg/dL creatinine and 99mg/dL urea. PCR of 0.98mg/dL. No changes in Urine II. Gasometrically, he revealed: pH 7.37 u. pH; pCO₂ 32mmHg; pO₂ 75mmHg; HCO₃ 18.5mmol/L; SatO₂ 94%.

The patient was admitted to the Internal Medicine Department for an etiological study and for compensation of ascites condition. He was submitted to a diagnostic and evacuation paracentesis and the ascitic fluid analysis revealed:

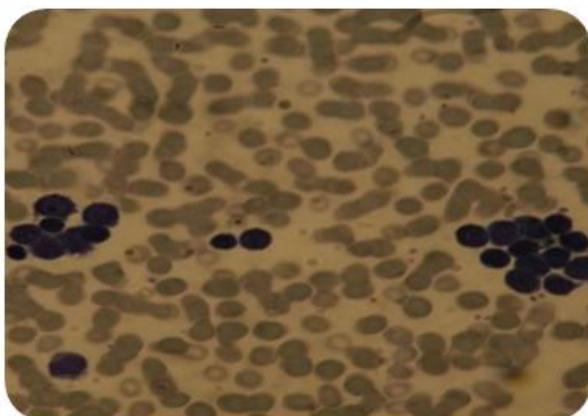


Fig-1: Cytological examination of ascitic fluid cells

- Cell count: 1025, with a 75% predominance of mononuclear cells;
- Glucose: 139 mg/dL, Albumin: 1.7 mg/dL and LDH 248 U/L → Serum-ascites albumin gradient: 1.4 → Transudate features;
- 30% hairy cells: CD19+, CD103+, CD11c+, LAIR1+, monoclonal kappa;
- 12% with Cd5 expression;
- Ascitic fluid compatible with infiltration by hairy cells;
- Anatomical-pathological report: "In the cytological examination, reactive mesothelial cells were observed in a background with abundant polymorphonuclear occasional lymphocytes. Negative for neoplastic cells".

In addition, he had no history of cirrhosis or analytical changes compatible with liver failure. Significant hypoalbuminemia or proteinuria was ruled out. Thyroid function was within the normal limits and serum electrophoresis with serum immunofixation did not detect monoclonal band.

Smears of peripheral blood revealed immunophenotyping compatible with hairy cell

leukaemia, with no apparent blasts, and presenting 10% of hairy cells.

The transthoracic echocardiography did not reveal changes that suggested right heart failure, making reference to the left ventricle (LV) with good global systolic function, dilated left atrium (LA), fibrosed aortic valve (AoV) and mitral valve (MV), as well as slight mitral failure (MF).

The liver Doppler revealed normal permeability and calibre of the portal trunk, thus, excluding portal vein thrombosis.

Thoracic-abdominal-pelvic computed tomography (CT) (no contrast due to chronic kidney disease (CKD)) showed mediastinal lymph nodes, slight hepatomegaly, and splenomegaly that had already been detected.

DISCUSSION

The ascites refers to the collection of fluid in the peritoneal cavity. Undeniably, the most common cause for ascites is portal hypertension related to liver cirrhosis. However, there are malignant and/or infectious causes that should always be ruled out [5]. The etiological study of ascites presents, once again, a challenge for the internists, since the differential diagnosis is extensive and many times depends on a detailed clinical history, where personal history, physical exam and complementary diagnostic exams acquire a high degree of importance. In the specific case presented, until the increase in abdominal size, the patient had always remained stable and autonomous despite being 90 years old. This aspect enabled maintaining an expectant approach. Given the inevitability of the progression of the disease, which was manifesting itself in an uncommon form, a low dose of chlorambucil was initiated in an attempt to delay its evolution. Despite regular evacuation paracentesis for the patient's comfort, the re-accumulation of ascitic fluid was quick, worsening concurrent dyspnoea and asthenia. In around 3 months of evolution, and particularly due to the enormous limitations of the treatment of the pathology at issue, the general status of the patient deteriorated, and he passed away in the Palliative Care Unit about 4 months after the first signs of refractory ascites. After the etiological investigation, it was admitted that the ascites was secondary to the hairy cell leukaemia variant due to the absence of any other identified cause for ascites. Although hairy cell leukaemia normally has an indolent course and does not develop with ascites, literature describes cases of hairy cell leukaemia as a cause for ascites, whose analysis of ascetic fluid identifies hairy cells, such as in this case. Thus, besides being before a pathology that is rare, the case refers a manifestation that is uncommon in the evolution of the disease.

CONCLUSION

Given a case like the one presented, where the etiology of the diagnosis is of exclusion, the laboratory assumes crucial importance, not only with the exclusion of other frequent causes (infectious, neoplastic), but also with the identification of hairy cells in the ascitic fluid that gear clinical suspicion towards the etiology of ascites. In the literature, despite the reference to cases of ascites as an uncommon manifestation of the evolution of hairy cell leukaemia, the truth is that the physiopathological mechanism is unknown and the relation between the two entities is ascertained by excluding other possible causes and the concurrent worsening of the hematologic disease [2, 4].

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