CASE REPORT

Coexistence of chylous ascites and thrombosis of the portal vein: case report and literature review

Coexistência de ascite quilosa e trombose da veia porta: relato de caso e revisão da literatura

Lennon da Costa Santos[1], Lucas Resende Lucinda[1], Guilherme Canabrava Rodrigues Silva[1], Anamaria Teixeira Gallo Rocha[1], Rosangela Teixeira[2], Luciana Diniz Silva[3]

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ABSTRACT

Chylous ascites (QA) is a rare condition, being characterized by the accumulation of lymph in the abdominal cavity. In adults, lymphomas constitute its most frequent cause; while cirrhosis and/or thrombosis of the portal vein are especially rare. This report presents a male patient, 36 years old, with chronic hepatitis C-related cirrhosis and alcoholism, 15 kg weight loss, and milky ascites with a predominance of triglycerides (1,500 mg/dL). The imaging methods identified the concomitance of thrombosis of the portal vein and cavernoma. The significant clinical improvement was obtained with the administration of total parenteral nutrition associated with octreotide. Alcohol abstinence was not achieved resulting in QA reappearance and deterioration of the clinical condition. The prognosis of QA in term of liver cirrhosis is bad. The treatment should be individualized according to the underlying clinical condition.

Key words: Chylous Ascites; Portal Vein; Venous Thrombosis; Alcoholism; Hepatitis C, Chronic; Liver Cirrhosis; Octreotide.

RESUMO

Ascite quilosa (AQ) é condição rara, sendo caracterizada pelo acúmulo de linfa na cavidade abdominal. Os linfomas constituem-se, em adultos, em sua causa mais frequente; enquanto cirrose e/ou trombose da veia porta são especialmente raras. Este relato apresenta paciente com 36 anos de idade, masculino, com cirrose associada à hepatite C crônica e alcoolismo, emagrecimento de 15 kg e ascite leitosa, com predominância de triglicérides (1.500 mg/dL). Os métodos de imagem identificaram a concomitância de trombose de veia porta e cavernoma. A melhora clínica significativa foi obtida com a administração de nutrição parenteral total associada ao octreotide. A abstinência ao álcool não foi conseguida e houve reaparecimento da AQ e deterioração da condição clínica. O prognóstico da AQ em vigência de cirrose hepática é ruim. O tratamento deve ser individualizado de acordo com a condição clínica subjacente.

Palavras-chave: *Ascite Quilosa; Veia Porta; Trombose Venosa; Cirrose Hepática; Alcoolismo; Hepatite C Crônica; Octreotida.*

[1] Medical School student at the Federal University of Minas Gerais – UFMG. Belo Horizonte, MG – Brazil.
[2] MD. PhD in Infectious Diseases and Tropical Medicine. Associate Professor in the Medical Clinical Department of the Medical School of UFMG. Belo Horizonte, MG – Brazil.
[3] MD. PhD in Gastroenterology. Associate Professor in the Medical Clinical Department of UFMG. Belo Horizonte, MG – Brazil.

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Institution:
Medical School at UFMG
Belo Horizonte, MG – Brazil

Corresponding Author:
Luciana Diniz Silva
E-mail: lucianadinizsilva@gmail.com

INTRODUCTION

Chylous ascites (QA) is a rare condition characterized by the accumulation of lymph in the abdominal cavity. Its physiopathology is complex and associated with the deregulation of the thoracic or abdominal lymphatic system[1,2] despite being commonly attributed to portal hypertension related to rupture and/or obstruction of lymph ducts.[3-17]

Elevated triglyceride concentrations in the peritoneal fluid are responsible for its milky appearance.[3]

In the West and East, and in developing countries, it is associated with malignant abdomen tumors, tuberculosis, and filariasis, respectively.[4] Chylous ascites associates to cirrhosis and/or thrombosis of the portal vein in 0.5 to 1.3%.[5-13]

This report describes a patient with cirrhosis related to chronic hepatitis C and alcoholism, with QA related to thrombosis of the portal vein.

DESCRIPTION OF THE CASE

A 36-year-old patient, male, admitted to the emergency room of the Risoleta Tolentino Neves Hospital with abdominal pain and distention, edema of lower limbs, weight loss of approximately 15 kg, and gastrointestinal bleeding. He reported the constant use of alcohol, about 600 g per day in the last eight years, and occupational exposure to blood and other human secretions. At admission, he presented systemic blood pressure of 90/60 mmHg, radial pulse of 92 bpm/min, hypo-colored mucous membranes, swelling of the lower limbs, axillary temperature of 38 ^{o}C, ascites, parotid hypertrophy, splenomegaly, muscle atrophy, abdomen collateral circulation, breath sounds decreased in lung bases; and no lymphadenomegalies.

The blood laboratory tests found: 9.2 g/dL hemoglobin; 3,200/µL leukocytes (68.7% neutrophils, 10.0% lymphocytes), 301,000/µL platelets; 137 mmol/L Na^{+};3.2 mmol/L K^{+}; 1.7 mmol/L Mg^{2+}; 41 mg/dL urea; 1.07 mg/dL creatinine; 91 mg/dL blood glucose; 54 U/L amylase; 31 U/L lipase; 108 U/L alkaline phosphatase; 178 U/L gamma glutamyl transferase; 57 U/L alanine aminotransferase; 100 U/L aspartate aminotransferase; 1.0 mg/dL total bilirubin; 153 U/L lactic dehydrogenase (LDH); 2.4 g/L albumin; 3.1 g/L globulin; 27.3 ng/dL ferritin; 10 mg/L C-reactive protein; 13.5 s prothrombin time; and polyclonal gammopathy revealed in the protein electrophoresis. The β_2-microglobulin dosage was 1.75 mg/L. Elevations of alpha-fetoprotein, CA-19-9, and CEA were not observed. The serology for the human immunodeficiency virus 1/2 and hepatitis B and VDRL was negative. The anti-HAV IgG and anti-HCV were positive [5.0 (normal < 1.0)], and the qualitative polymerase chain reaction (PCR) confirmed the presence of hepatitis C virus RNA in the plasma.

The paracentesis drained eight liters of a milky liquid (Figure 1), which revealed in the biochemical analysis: 25/mm^3 leukocytes (100.0% mononuclear); 115 mg/dL glucose; 10.0 U/L amylase; 79 U/L LDH; triglycerides1,500 mg/dL; adenosine deaminase less than 40 U/L; 1.4 g/dL total protein; 0.6 g/dL albumin, soroascite albumin gradient of 1.8; and acid alcohol bacilli resistance fast staining by Ziehl-Neelsen (AABR) and culture, as well as search for other bacteria and fungi.

Figure 1 - Aspect of ascetic fluid: Ascetic fluid rich in triglycerides (1,500 mg/dL) with an appearance similar to milk.

Alterations in the chest x-ray or echocardiogram were not identified. Abdomen magnetic resonance imaging (MRI) and ultrasound examination showed no malignancy or adenomegalies, being observed signs of liver cirrhosis with portal hypertension and thrombosis of the portal and splenic veins (Figure 2), cavernoma in the portal vein, and accentuated splenomegaly and ascites. The high digestive endoscopy showed gastroesophageal varicose veins of thin gauge without signs of bleeding.

Neoplasia and lymphadenomegaly were not observed in the chest computed tomography, however, an in caverna image in the anterior segment of the upper left lobe was detected. The histological analysis of a lung biopsy, guided by fibro bronchoscopy, showed no signs of malignancy or tuberculosis. The Ziehl-Neelsen staining and culture in this analysis were negative. The tuberculin test and multiple analyses of sputum were negative. The diagnosis of chylous ascites was established based on these findings and probably due to portal hypertension and portal vein thrombosis.

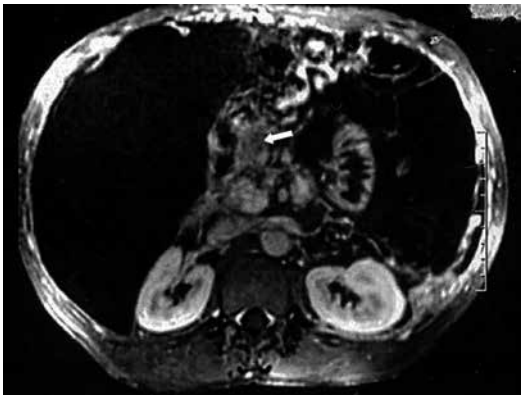

Figure 2 - Abdomen MRI showing liver cirrhosis with signs of portal hypertension and portal vein thrombosis.

The patient was treated with diuretics, low sodium diet, with supplementation with medium chain triglycerides. However, he evolved with refractoriness and recurrence of QA. Subcutaneous octreotide (three times a day) and total parenteral nutrition were initiated after four weeks of follow-up. A reduction of abdominal distension was observed after six weeks of therapy with a remarkable reduction of symptoms in the following three weeks. The patient was discharged from the hospital with the proposal of alcohol abstinence and behavioral change for a more appropriate and healthy life style.

He could not abstain from alcohol and after one year of monitoring, QA resurfaced, and a gradual deterioration of his health conditions set in. The hospitalizations became recurrent due to ascites, with repeated paracenteses and milky fluid drainage.

DISCUSSION

The evolution of QA is described in this article as the result of cirrhosis and portal vein thrombosis characterized by progressive painful abdominal distension during three weeks, accompanied by weight loss, edema, fever, gastrointestinal bleeding, and anemia in addition to stigmas of chronic liver disease on the physical examination. Risk factors that predispose to cirrhosis were identified such as an occupational accident with contaminated blood and high consumption of alcohol.

The decisive role of paracentesis for the diagnosis and patient monitoring must be emphasized. The milky appearance of the ascites fluid, rich in triglycerides (1,500 mg/dL), contributed to the establishment of the QA diagnosis; [1] in addition, other laboratory and microbiological tests were carried out on the drained liquid off of the peritoneal cavity, which allowed the exclusion of other diseases, especially tuberculous peritonitis.

Tuberculosis remains an important cause of illness and death in Brazil.[18] The prevalence of tuberculosis is higher in poorer areas, particularly associated with alcohol consumption.[19] In this case, tuberculosis needs to be excluded. The QA has been reported in some clinical situations,[1] especially in adults, due to: neoplasias, especially lymphomas.[20] Cirrhosis and/or thrombosis of the portal vein are rare causes.[5-13] The transformation of yellow and transparent aspect of the ascites fluid in chylous is associated with a worsen prognosis.[6]

The diagnosis of QA secondary to cirrhosis and portal vein thrombosis was made, in this case, although rare, based on the lack of evidence of malignancy and tuberculosis. The aid of complementary examinations, such as abdomen MRI revealed cavernoma of the portal vein, characterized by complex local network of collateral veins.

The presence of compensatory mechanisms is essential for maintaining the hepatic perfusion in spite of cirrhosis and portal vein thrombosis.[15] Thus, a complex network of collateral veins was formed to bypass the thrombosed segment.[10] As a result, portal hypertension developed, which caused excess lymphatic flow; its overload spurred the rupture of lymphatic vessels and lymph stasis and exudation to the peritoneal space.[4]

The QA approach requires a hyperproteic and hypolipidic diet, which reduces the production and flow of the lymph; and in patients with cirrhosis, low sodium diet and use of diuretics. In refractory ascites, the rapid reduction of lymph flow and total parenteral nutrition is used.[4,9-10]

Somatostatin and its analogues have been effectively used in the treatment of QA patients secondary to the rare Yellow Nail Syndrome and lymphatic escape caused by abdominal and thoracic surgeries.[21-24] The use of octreotide for QA in patients with cirrhosis is less frequently reported.[9-10,25] Therefore, the possible improvement of the clinical picture is related to the effect of a rapid reduction in portal pressure.

Hence, the conduct was based on the combination of various therapeutic techniques. There are no conclusive guidelines for monitoring QA. Several studies emphasize the need for individualized treatment according to the underlying clinical conditions.[4] In QA caused by liver cirrhosis, numerous evidences indicate that the reduction of portal pressure

is of fundamental importance. The administration of octreotide appears to be effective when there is the coexistence of QA and portal vein thrombosis in patients with cirrhosis.[9-10,25]

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