CHYLOTHORAX IN HAIRY CELL LEUKEMIA

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Abstract
A case of postoperative left chylothorax in a 43-year-old black woman with hairy cell leukemia is reported. First submitted to pleural drainage, she was successfully treated with a combination of chemotherapy and elemental enteral diet enriched with medium-chain triglycerides.

Case Report
A 43-year-old black woman came to our hospital on May 1996, complaining of continuous dyspnea for a week. She also related lack of appetite, asthenia, and loss of about 10% of her usual body weight during the last 3 months. As a consequence of hairy cell leukemia, she had been submitted to splenectomy, without complications, 3 weeks before. She was in regular use of oral estrogen, because of an hysterectomy four years before. She denied use of tobacco or alcohol beverages. Physical examination showed pale (++) mucosa and no peripheral edema. Anthropometry showed weight 47.3 kg, height 1.54 m, body mass index (BMI) 19.94 kg/m² and moderate fat and lean corporal mass deficit. Axillar temperature 36.8 °C. The heart was normal, with 120 bpm. Blood pressure 100/60 mmHg. There was physical evidence of a voluminous left pleural effusion. The liver and lymph nodes were normal on palpation. Erythrocyte count 4.17 x 10⁶/mm³, hemoglobin 11.9 g/dl, hematocrit 38.1%, MCV 91 fl, MCH 28.4 pg, MCHC 31.2 g/dl, moderate anisopoikilocytosis, reticuloctyes 70 000/mm³, leucocyte count 8 10³/mm³ (neutrophils 50%, lymphocytes 35%, monocytes 6%), lymphocytes with abundant cytoplasm and cytoplasmic projections, platelets 511 000/mm³. Erythrocyte sedimentation rate 60 mm. Glucose 143 mg/dl, urea 17 mg/dl, creatinine 0.8 mg/dl, uric acid 2.6 mg/dl, sodium 145 mEq/l, potassium 4.7 mEq/l, chloride 107 mEq/l, calcium 9.7 mg/dl, phosphorous 4.4 mg/dl, total bilirubin 0.35 mg/dl, AST 26 U/l, ALT 24 U/l, LDH 485 U/l, γGT 125 U/l, alkaline phosphatase 485

Chylothorax is the effusion of chyle in pleural cavity due to lymphatic drainage obstruction or thoracic duct damage. This infrequent condition is more often seen on the right hemithorax¹ and diagnosis is established when pleural fluid contains chylomicrons and high levels of triglycerides (> 110 mg/dl). The causes of chylothorax are blunt or open thoracic trauma, abdominal or thoracic surgery, congenital lymphatic anomalies, subclavian and superior cava vein thromboses², inflammatory processes or malignant neoplasia, in special lymphomas³. After repetitive thoracocenteses or continuous drainage, lymphocytes, fat and protein body losses are observed, which rapidly impair the patient’s nutritional status⁴. Clinical management may be curative, and includes the regimen of nothing per os and total parenteral nutrition (TPN). Case studies have also suggested benefits of a low fat diet, sometimes enriched with medium-chain triglycerides (MCT), and enteral elemental diet⁵.

We report a case of a female with hairy cell leukemia (HCL), a rare disease of B lymphocytes (1-2% of all leukemias) occurring more frequently in men (4:1). Our patient developed left chylothorax three weeks after splenectomy; first submitted to pleural drainage, she was successfully treated with a combination of chemotherapy and elemental enteral diet enriched with MCT.

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U/l, amylase 104 U/l, mucoprotein 129 mg/dl, tyrosine 8.6 mg/dl, fibrinogen 5.7 g/l, iron 34 µg/dl, ferritin 65.9 ng/ml, LIBC 211 µg/dl, TIBC 245 µg/dl, albumin 3.6 g/dl, globulin 4.0 g/dl, cholesterol 160 mg/dl, triglycerides 118 mg/dl, HDL 39 mg/dl, VLDL 24 mg/dl, LDL 97 mg/dl. Alphaglycoprotein reagent (153), IgG 1 270, IgA 284, IgM 322. Complement proteins: C3 122 µg/ml, C4 37.1 µg/ml. Toxoplasmosis (IFI) IgG reagent 1:16 and IgM non-reagent. Chagas’ disease serologic reactions negative. HIV serologic tests negative (cutoff: 0.142 ng/ml and patient: 0.044 ng/ml). Radiography of thorax confirmed normal heart size and voluminous left pleural effusion (Figure 1A). Biopsy of the right iliac crest showed a hypocellular bone marrow with discrete plasmocytosis associated with lymphocytic interstitial infiltration, besides fibrosis and hemossiderin deposits. Bone marrow aspirate material showed global moderate cellular paucity, and many atypical spiculate lymphocytes, characteristic of hairy cells (Figure 2). Although HCL immunophenotype was not established, the presence of tartrate-resistant acid phosphatase reaction and the immune histology (DBA 44) confirmed the HCL diagnosis.

In order to alleviate dyspnea, three thoracocenteses were performed, draining a total of 5 450 ml of a thick, chocolate-milk colored liquid, pH 8.0, specific gravity 1035, cells count 900/mm³ (lymphocytes 51%, neutrophils 18%, mesothelial 31%), erythrocytes 26 000/mm³, glucose 89 mg/dl, protein 5.4 g/l, LDH 823 U/l, cholesterol 72 mg/dl, triglycerides 1107 mg/dl. Bacterioscopy (Gram) and all laboratory tests to detect neoplastic cells, fungus and alcohol-acid fast bacteria were negative. After the last thoracocentesis, a computerized tomography (CT) demonstrated thoracic duct of normal diameter and absence of intrathoracic masses.

As pleural effusion rapidly accumulated after drainage, a nasoenteral elemental diet containing 1 870 kcal and 46 g of protein was supplied by continuous infusion pump (42 ml/h), besides intravenous lipid emulsion 10%, 500 ml twice a week. Because cladribine is considered the drug of choice in HCL, a single continuous intravenous infusion course of this adenosine deaminase-resistant purine analog (0.09 mg/kg/day) was used for 7 days. The patient had an uneventful clinical course and, after seven days of elemental enteral nutrition, low fat, MCT enriched oral diet started. The control radiographic study confirmed total chylothorax disappearance (Figure 1B). MCT of diet was gradually changed for soya oil and, at discharge, she was receiving free diet without any problem. After one year of hematological outpatient follow-up there was no chylothorax relapse.
Discussion

To our knowledge, the association of chylothorax with hairy cell leukemia has not been previously reported in the literature. Bouroncle (1987), in a large series of 116 HCL patients, did not find any case of chylothorax among other rare associated conditions, including protein losing enteropathy, ascites and massive pleural effusion. Pleural fluid analysis showed high triglyceride levels (1.107 mg/dl) and a large number of lymphocytes, which confirmed the chylothorax diagnosis.

In the present case, chylothorax etiology is not clear, but there are two main possibilities: splenectomy three weeks before admission, and/or lymphatic abnormalities secondary to leukemia. Splenectomy has not been reported among the etiologies of chylothorax; nevertheless, left pleural chylosus effusion was described after nephrectomy and following accidental or surgical abdominal trauma above the sixth thoracic vertebra level. Although CT does not substitute lymphangiography, our data showed thoracic duct of normal diameter and no intrathoracic masses, suggesting absence of lymphatic flow interruption, as may occur in lymphomas. Moreover, lymphadenomegalies rarely occur in HCL.

Excessive chyle drainage causes losses of large amounts of fat, protein, electrolytes, and lymphocytes. Nutritional therapy is essential in chylothorax management to prevent malnutrition and to reduce chyle production, decreasing lymph leaking into pleural space. In this case, the patient was in an accelerated development of malnutrition, evident by rapid loss of body weight associated with anorexia, low food intake and a daily pleural lymph loss of 780 ml, which contains about 42 g of albumin.

Surgical ligation of the thoracic duct is sometimes indicated after 2-3 weeks of conservative treatment with chest tube drainage and nutritional therapy. TPN is an efficient way to reduce lymphatic fistula flow, but could be harmful to a patient with immunodepression, predisposed to venous-catheter infection and sepsis. Oral intake is not the therapy of choice in chylothorax, because it often contains fat which increases chyle production. Monomeric or elemental enteral diet consists of an admixture of aminoacids, sucrose, vitamins and minerals. It does not contain triglycerides, is readily absorbed even in cases of intestinal atrophy, and protect the intestinal mucosa against microbial translocation and the risk of gut-origin sepsis. MCT are directly absorbed in the portal vein system, decrease the chyle flow, and reduce the thoracic duct pressure, minimizing lymph leaking, and eventually promoting fistula closing.

References