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Scientific letter

Portal Hypertension: An Uncommon Cause of Chylothorax and an Atypical Association With Systemic Sclerosis

Hipertensión portal: una causa infrecuente de quilotórax y una asociación atípica con la esclerosis sistémica

Dear Editor,

We report the case of a 52-year-old male with a medical history of systemic sclerosis (SSc) and portal hypertension, associated with esophageal varices and bleeding. He has no history of alcohol consumption or other toxic substances, and liver disease was ruled out by liver biopsy.

He attended the respiratory medicine department for a study of dyspnea. An initial suspicion of interstitial lung disease (ILD) associated with SSc was finally ruled out. Chest X-rays and ultrasound revealed right pleural effusion with no lung parenchymal involvement (Fig. 1a and b). Subsequent thoracentesis was carried out, yielding milky-appearing pleural fluid (Fig. 1c). Laboratory studies confirmed transudative chylothorax (protein 1.65 g/dl, LDH 91 U/L, triglycerides 229 mg/dl). Cytology results ruled out malignancy. The patient had no history of trauma, recent surgery or known neoplastic processes. Several tests were conducted to rule out potential causes. A computed tomography scan of the chest and abdomen revealed splenomegaly and collateral circulation due to pre-existing portal hypertension, previously confirmed by a hemodynamic study (wedged hepatic venous pressure (WHVP) 17.5 mmHg, free hepatic venous pressure (FHVP) 8 mmHg, hepatic venous pressure gradient (HVPG) 9.5 mmHg). Minimal ascites was noted, but no other anomalies were detected (Fig. 1d–f). Echocardiography showed normal biventricular function with indirect signs of pulmonary hypertension. Right heart catheterization confirmed precapillary pulmonary hypertension group 1 (mean pulmonary arterial pressure (mPAP) 87 mmHg, pulmonary arterial wedge pressure (PAWP) 5 mmHg, pulmonary vascular resistance (PVR) 23 WU). Laboratory tests for anti-mitochondrial, anti-smooth muscle cell and anti-LKM antibodies were negative (ruling out primary biliary cholangitis and autoimmune hepatitis), antinuclear and anti-Sc70 antibodies were positive, hepatic serologies nega-

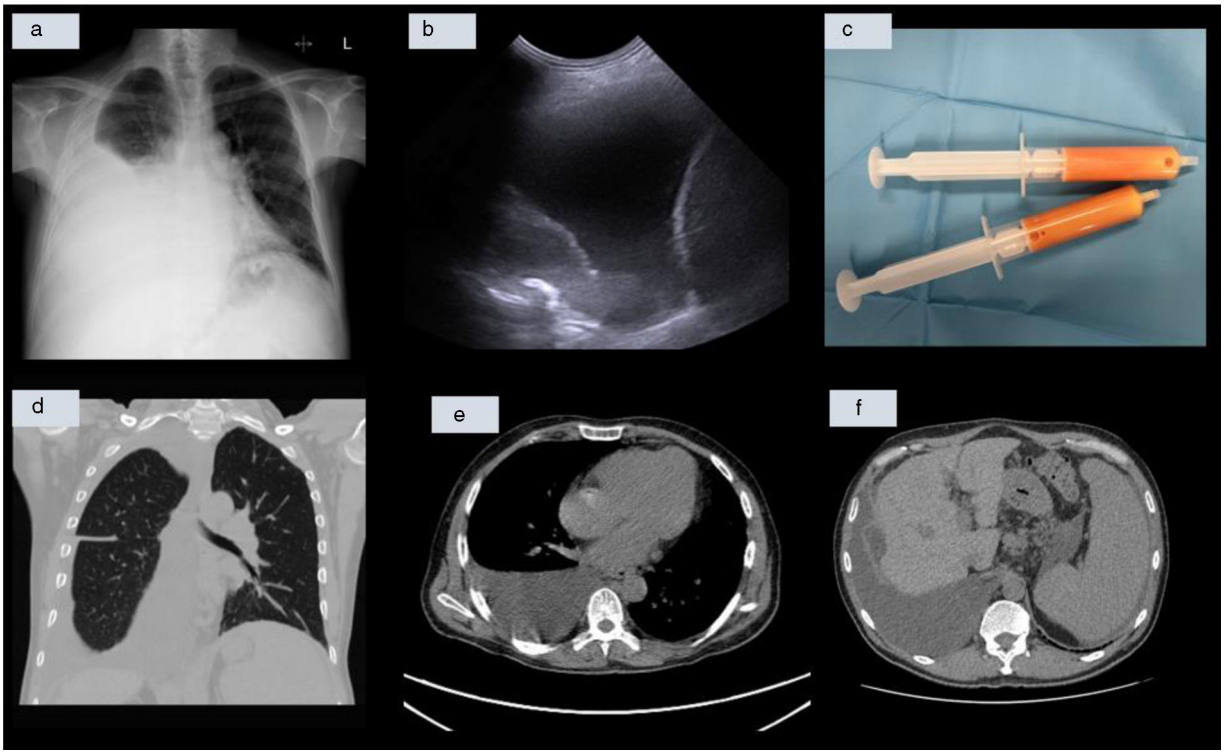

Fig. 1. (a) Chest X-rays showing right pleural effusion; (b) chest ultrasound revealing anechoic pleural effusion; (c) milky-appearing pleural fluid; (d and e) chest CT scan with pleural effusion with no lung parenchymal involvement; no other anomalies were detected; (f) abdominal computed tomography scan with splenomegaly and ascites.

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tive, and the blood cell count was normal. Ultimately, and despite the exceptional nature of this manifestation, the chylothorax was attributed to portal hypertension and managed with dietary modifications alone.

This case is of interest given the rarity of transudative chylothorax associated with portal hypertension secondary to SSc, which, in itself, is exceptionally uncommon.[1,2]

Chylothorax manifests as an accumulation of chyle in the pleural space, typically as a result of thoracic duct disruption or the passage of chyle from the abdominal cavity. Etiologies can be classified as traumatic, including postsurgical, in up 50% of the cases; and non-traumatic, similarly in 50% of cases, most frequently lymphoproliferative malignancies. Non-malignant causes include liver cirrhosis, heart failure, nephrotic syndrome, tuberculosis, sarcoidosis, lupus erythematosus and congenital disorders of the lymphatic system, among others. All of these were ruled out in our patient. When the underlying cause remains unidentified, chylothorax is considered idiopathic, as occurs in up to 6%–14% of cases.[3] Although rare, cases of chylothorax resulting from portal hypertension due to cirrhosis have been reported.[4] Our patient, notably, did not have liver disease.

Chylothorax is typically diagnosed through the study of pleural fluid. The fluid is often milky due to its high triglyceride content, although this characteristic is non-specific, as it can also appear in cholesterol effusion or empyema. In patients following a low-fat or fat-free diet, the fluid may appear sanguineous or serous. Its triglyceride concentration is typically >110 mg/dl, and the identification of chylomicrons by lipoprotein electrophoresis confirms the diagnosis. Most cases are exudative, with a predominance of lymphocytes. However, our case was a transudative chylothorax, less common and related in the majority of cases with liver cirrhosis. Other causes of transudative chylothorax reported in the literature include congestive heart failure, superior vena cava obstruction, pulmonary hypertension, and amyloidosis.[5,6] The management of chylothorax depends on its underlying cause and typically involves dietary modifications and pleural space drainage. Surgical intervention may be considered in cases of persistent high output.[7,8]

In summary, chylothorax is a rare cause of pleural effusion. Its association with portal hypertension (unrelated to liver disease) due to systemic sclerosis is unusual.

Informed consent

The patient's consent for publication has been obtained.

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Conflicts of interest

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