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Letters to the Editor

Partial Anomalous Pulmonary Venous Drainage and Mayer-Rokitansky-Küster-Hauser Syndrome☆

Drenaje venoso pulmonar anómalo parcial y síndrome de Mayer-Rokitansky-Kuster-Hauser

To the Editor:

Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome is characterized by congenital agenesis of the uterus and vagina in women with normal ovaries, phenotype and karyotype.[1,3] The associated lung malformations described are practically anecdotal.[1–4] We report a case of associated partial anomalous pulmonary venous drainage.

The patient is a 19-year-old woman with MRKH, who referred frequent colds and presented low-grade fever, purulent expectoration and dyspnea with minimal exertion. Physical examination was normal, except for mild tachypnea and bibasilar inspiratory rales. The work-up presented leukocytosis with neutrophilia and the arterial blood gas showed pCO_2 39 and pO_2 57. Chest radiograph and electrocardiogram were normal. Thoracic computed tomography (CT) and magnetic resonance imaging (MRI) revealed partial anomalous pulmonary venous drainage: the vein of the upper left lobe was draining into the innominate vein through the vertical vein (Figs. 1 and 2). Transthoracic echocardiogram with contrast ruled out intracardiac communications as well as overload of the right cavities.

MRKH is a rare malformation (incidence 1/4000 living newborns) characterized by congenital agenesis of the uterus and vagina in women with normal ovaries, secondary sex characteristics and karyotype.[1–4] The etiopathogeny lies in a lack of development of the Müllerian ducts, with the consequent alteration in the structures derived from them.[1] The diagnosis is based on primary amenorrhea and the inability to allow for vaginal penetration, confirmed by ultrasound and MRI.[1,4] It is divided into two types: typical or I (agenesis of the uterus and vagina) and atypical or II, associated with other malformations (renal, skeletal, digital and cardiac, as well as deafness).[1–4] The associated pulmonary malformations that have been published are practically anecdotal, including a case of pulmonary hypoplasia[4] and a case of total anomalous pulmonary venous drainage.[3] We describe a case of partial anomalous pulmonary venous drainage where the vein of the upper left lobe drains into the innominate vein through the vertical vein. Anomalous pulmonary venous drainage is produced when the pulmonary venous blood from one or more veins flows directly into the right auricle or in the systemic veins. It may be total (4 anomalous veins) or partial (1–3 anomalous veins). In both types, a left-to-right extracardiac shunt is produced, causing an increase in bloodflow with the consequent overload in the right auricle and ventricle, as well as in the pulmonary artery and veins. This hyperflow can cause heart failure and pulmonary hypertension. The hemodynamic overload depends on the number of veins with anomalous connection, the location of the drainage and whether it is associated with intraauricular communication. Thus, the patients can be asymptomatic, present frequent colds—as in our case—or

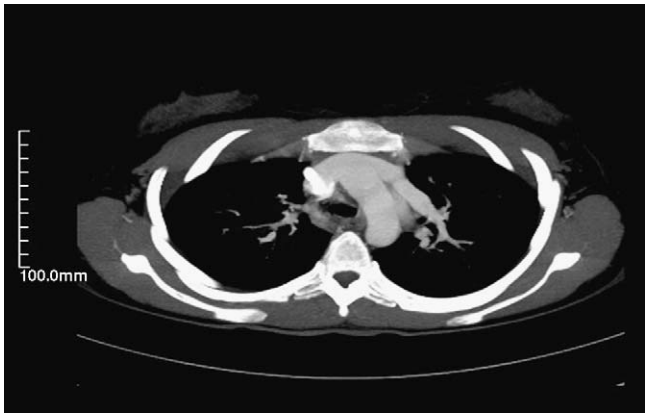

Fig. 1. Partial anomalous pulmonary venous drainage.

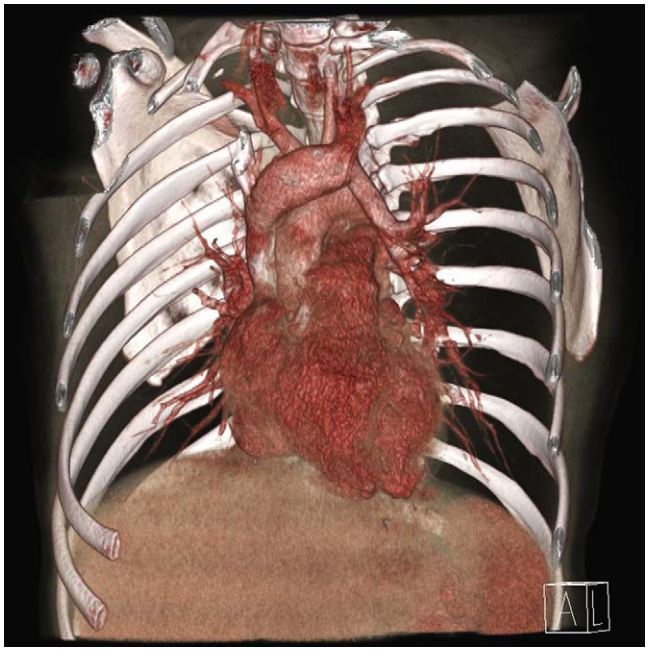

Fig. 2. The vein of the left upper lobe drains into the innominate vein through the vertical vein.

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present heart failure and pulmonary hypertension. The anomalous vein on occasion can be seen on simple radiography, confirming the diagnosis by CT and MRI, and resorting to cardiac catheterization when given the suspicion of pulmonary hypertension.

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Diffuse Pulmonary Lymphangiomatosis With Mediastinal Affectation☆

Linfangiomatosis pulmonar difusa con afectación mediastínica

Dear Editor:

Lymphangiomatosis is an uncommon systemic disease characterized by a marked proliferation and dilatation of the lymphatic vessels.[1] It can appear as either localized or diffuse affectation, in which case it is known as diffuse pulmonary lymphangiomatosis (DPL). It may affect newborns and young adults. Although it is a benign disease of unknown etiology and pathogeny, it presents a high rate of relapse and a poor prognosis.

We present the clinical case of a 55-year-old male patient, ex-smoker with a history of hypertension, dyslipidemia and COPD with bronchodilator treatment. He came to the emergency unit due to dyspnea with minimal exertion that had been evolving over the period of a month. The basic analytical data were normal; chest radiograph revealed basal bronchiectasis, and spirometry presented an obstructive pattern. On computed tomography (CT), multiple mediastinal lymphadenopathies were observed as well as perihilar interstitial affectation (Fig. 1A) and pericardial effusion. Bronchoscopy revealed diffuse thickening of the bronchial mucosa and widening of the main and bronchial carinas. The mediastinal affectation was studied using mediastinoscopy, where anthracotic-looking lymphadenopathies and fatty nodules were observed; their biopsies presented lymphorrhea. The study of the liquid drained from the mediastinum confirmed that it was chylous with no malignant cellularity; the microscopic study of the lymphadenopathies was not conclusive. Afterwards, a lympho-gammagraphy was performed, where adequate lymphatic migration was seen without signs of leakage at the level of the thoracic duct. Given the lack of diagnosis and persistence of the dyspnea, pulmonary and mediastinal biopsies were taken by means of video-assisted thoracoscopy. The anatomopathologic study revealed proliferation of dilated capillary vessels in the pleura and septa and mediastinal adipose tissue with proliferation of anastomosed lymphatic vessels and lymphoid accumulations, leading to the definitive diagnosis of DPL. After successive episodes of dyspnea with minimal exertion secondary to pleural effusion (chylothorax), surgical treatment was indicated. Video-assisted thoracoscopy showed evidence of the characteristic proliferation and dilation of the lymphatic vessels and lymphorrhea in the lung parenchyma and mediastinum (Fig. 1B). Ligation of the thoracic duct and pleurodesis was carried out. Post-operative evolution was favorable, and the patient was discharged 10 days later.

DPL is an uncommon lymphatic alteration that appears in children and young adults, while it is quite exceptional in patients over the age of 40.[1] The most frequent symptom is dyspnea. It may be erroneously diagnosed as asthma responding to bronchodilator treatment, which was the reason for the late diagnosis in our case, previously diagnosed with COPD with bronchodilator treatment. It is associated with pleural effusions, other lymphatic abnormal-

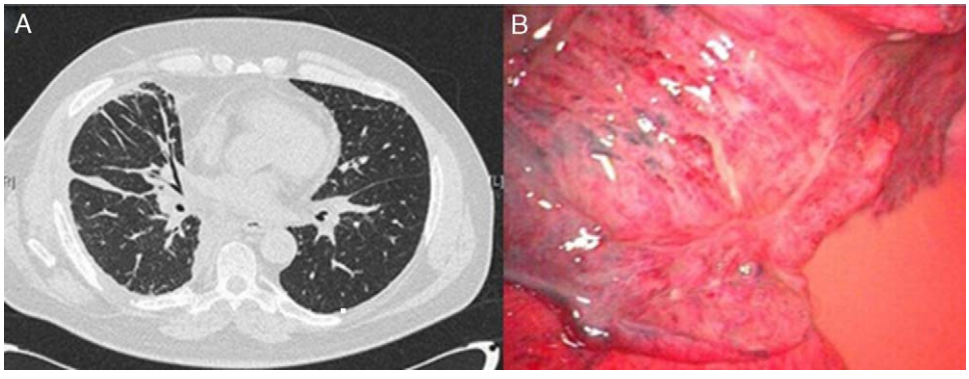

Fig. 1. (A) Perihilar interstitial affectation and mediastinal adenomegalies; (B) thoracoscopic image showing the dilatation of the subpleural lymphatic vessels, lymphorrhea and the resulting chylothorax.

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