Unilateral Congenital Atresia in Adults. A Case Report and Review of the Literature

Atresia congénita unilateral de venas pulmonares en adultos. Descripción de un caso y revisión de la literatura

To the Editor,

Congenital unilateral pulmonary vein atresia with no other cardiac abnormalities is a very rare entity. It usually presents in childhood or adolescence as recurrent episodes of lung infection or hemoptysis, and is an exceptional finding in adults. We have only found 11 cases in the literature to date (including our own).

We present the case of a 43-year-old man, who we examined after admission for pneumonia. His medical history included type 1 diabetes and congenital right pulmonary venous atresia diagnosed in childhood, with no subsequent follow-up. This was the third episode of pneumonia in the right upper lobe in 2 years. He also reported small-volume hemoptysis daily, and dyspnea on major exertion. Decreased breath sounds were observed throughout the right hemithorax on auscultation, with persistent wheezing in the anterior axillary line.

Chest radiograph showed a resolving consolidation together with decreased right lung volume with a mediastinal shift to that side. The computed tomography (CT) scan is shown in Fig. 1. Bronchoscopy was performed, with the following findings: absence of right upper lobe bronchus; in its place was the opening of 2 accessory bronchi that prevented the bronchoscope from advancing. The opening of the apical and paracardiac bronchus of the right lower lobe could not be seen either. Spirometry revealed a moderately restrictive pattern, while the echocardiogram showed mild aortic stenosis with no pulmonary hypertension findings.

Finally, an angiography was performed. This confirmed all the findings described in the CT and also showed that part of the collateral system formed to supply the right lung came from an inferior phrenic artery (Fig. 1).

After the study was completed, the case was discussed in a meeting with the thoracic surgery and vascular surgery departments, where it was decided that the most appropriate therapeutic option was pneumonectomy.

Unilateral pulmonary vein atresia occurs due to failure of incorporation of the common pulmonary vein into the left atrium. It is associated with other cardiac abnormalities in up to 50% of cases, and may occur in either lung.


citation

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References

The clinical spectrum ranges from asymptomatic patients to the development of pulmonary hypertension and death. Diagnosis is confirmed by pulmonary angiography. However, modern techniques such as CT scan or magnetic resonance imaging together with symptoms consistent with the condition may now be sufficient to make the diagnosis.

There are 3 therapeutic options proposed in the literature. Pneumonectomy has been shown to control symptoms, and to prevent pulmonary hypertension. Another, less aggressive, therapeutic option is coil embolization of the systemic arterial collateral vessels. In the article by Heyneman et al., this procedure controlled hemoptysis in one of the patients, who subsequently remained asymptomatic. The third option, in cases of asymptomatic patients or those with few symptoms, is follow-up, focusing particularly on the early detection of pulmonary hypertension.

References


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Isolated Unilateral Pulmonary Vein Atresia in Adults

Atresia aislada unilateral de venas pulmonares en el adulto

To the Editor,

We present the case of a 19-year-old woman, smoker (8 pack-years), and regular cannabis user, with a history of severe bronchitis in childhood and frequent respiratory infections. She presented for episodes of coughing with hemoptoic expectoration since she was 15-years-old. The episodes were initially sporadic, but had become more frequent in recent months, sometimes occurring every 48 h. On examination, she had decreased breath sounds in the right hemithorax, and acropathy. The chest radiograph showed loss of volume in the right lung and attenuation of the right hilum. The right pulmonary veins could not be identified on computed tomography (CT), and a diffuse increase in density was observed in the affected lung. Cardiac magnetic resonance imaging (MRI) ruled out associated heart disease, and the echocardiography ruled out pulmonary hypertension. Pulmonary arteriography was performed (Fig. 1A and B), in which hypoplasia of the right pulmonary artery and absence of right pulmonary venous return were observed. Arteriography of the thoracic artery (Fig. 1C) showed right bronchial artery hypertrophy originating in the descending aorta. Lung function test results were: FEV1: 2.040 (75.5%), FVC: 2.800 (90.1%) and DlCo/SB: 69.3%. Varices were observed in the distal trachea and start of the right main bronchus on fiber optic bronchoscopy. Considering the previous findings, we opted for surgical treatment, specifically, right pneumonectomy. The immediate post-operative period was uneventful, and the patient is currently asymptomatic.

Fig. 1. Pulmonary arteriography showing hypoplasia of the right pulmonary artery, providing little perfusion, almost entirely limited to the upper lobe (A), in which right venous drainage cannot be seen (B). Arteriography of the thoracic aorta showing an irregular, winding bronchial artery with origin in the anterior superior side of the descending thoracic aorta (C).

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