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Unilateral Agenesis of the Pulmonary Artery (AUPA) Associated with Chronic Pulmonary Thromboembolism

Agenesia Unilateral de la Arteria Pulmonar (AUAP) Asociada a Tromboembolia Pulmonar Crónica

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CASE REPORT

24-year-old male patient who is a university student and practices mixed martial arts. He has no personal history of pathological conditions, but reports a surgery for an unspecified, apparently benign mass on his lower lip 15 years ago. He comes to the outpatient service due to a clinical condition characterized by cough and mild hemoptysis of one month of evolution, dyspnea upon moderate exertion that progresses to minimal exertion, and unquantified weight loss. No fever or night sweats.

Physical examination: blood pressure, 100/70 mmHg; heart rate, 103 beats per minute; respiratory rate, 16 breaths per minute, with 95% oxygen saturation (FiO_2 0.21). On auscultation, diminished vesicular sounds were noted at the left apex along with ipsilateral pectoriloquy. Right lung field sounds were normal.

Due to epidemiological history of residing in a region with high incidence of tuberculosis, two sputum samples were taken for bacilloscopy, both of which were negative. The simple CT scan of the chest showed decreased lung attenuation in the left field, predominantly in the upper lobe and lingula, as well as a hypodense image in the right pulmonary artery, indicative of pulmonary thromboembolism.

The contrast-enhanced CT angiography of the chest showed absence of the left pulmonary artery at the bifurcation level, and several collateral branches originating from a tortuous descending aorta with multiple ramifications (Figures 1 and 2). Chronic thrombus was also noted in the main branch of the right pulmonary artery (Figure 3).

The electrocardiogram showed sinus rhythm, pulmonary P wave, and signs of right ventricle hypertrophy and overload.

The transthoracic echocardiogram showed dilation of the right ventricle, dilation of the pulmonary trunk, and a systolic pulmonary artery pressure (sPAP) of 40 mmHg. No intracardiac shunts or masses were observed.

Lower limb Doppler ultrasound was performed with compressible vessels, without images inside, doppler flow present, negative for deep vein thrombosis.

Laboratory studies reported normal blood count with platelets at 179,000; D-dimer: 780; uric acid: 7.4; non-reactive VDRL and HIV; PT: 16.6; INR: 1.53; PTT: 35.7; negative C-ANCA and P-ANCA; negative ANA; C3: 116; C4: 25; IgG cardiolipin antibodies: 24.98; IgM cardiolipin antibodies: 12.27;

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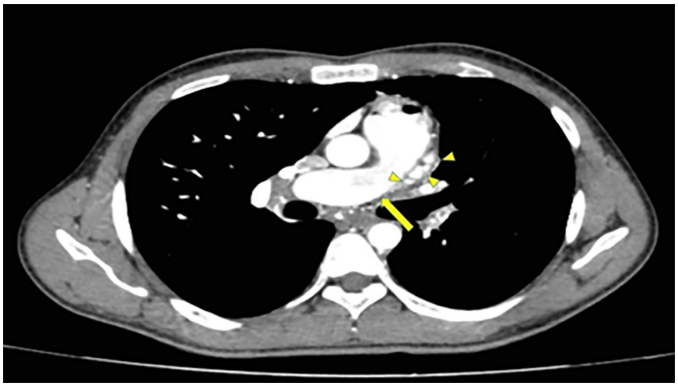

Figure 1. Axial section of contrast-enhanced pulmonary angiotomography: Absence of left pulmonary artery at the bifurcation level (*arrow*). Multiple collateral branches coming from the descending aorta artery (*arrowheads*).

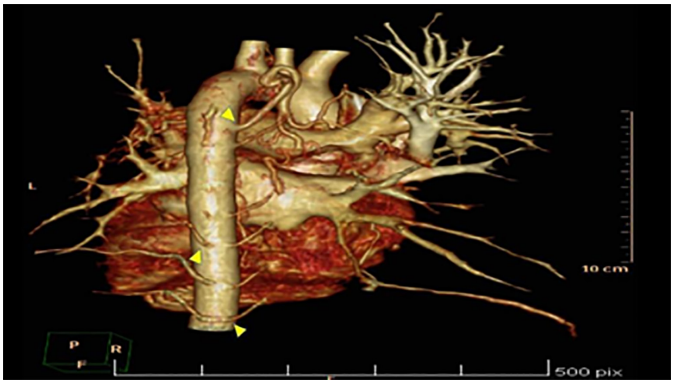

Figure 2. Three-dimensional cardiac reconstruction: Multiple collateral branches coming from the descending aorta artery (*arrowheads*).

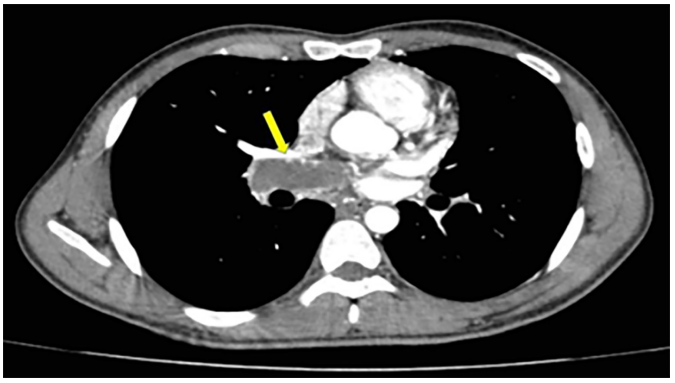

Figure 3. Axial section of contrast-enhanced pulmonary angiotomography: Presence of chronic thrombus in right pulmonary artery (*arrow*).

rheumatoid factor: 20.80; homocysteine 7.01; negative direct Coombs test; negative tumor markers; IgG and IgM for cytomegalovirus, toxoplasma, rubella, herpes negative; negative hepatitis C; CPK: 89; CKMB: 38.80; fibrinogen and coagulation factors within normal parameters.

It was concluded that the unilateral pulmonary artery agenesis in this patient was not related to any cardiac anomalies, and the pulmonary thromboembolism was idiopathic. The patient showed remission of hemoptysis, so anticoagulation with rivaroxaban was initiated.

Unilateral agenesis of the pulmonary artery is a rare malformation with a prevalence of one in every two hundred to three hundred thousand young adults. Due to its low frequency, it constitutes a diagnostic challenge and is often underdiagnosed in pediatric age.[1]

It occurs as a consequence of intrauterine involution of the ipsilateral sixth aortic arch, causing a failure in the connection of this arch with the pulmonary trunk, thus conditioning the absence of the proximal portion of the right or left pulmonary artery.[2]

Pulmonary artery agenesis is more prevalent in the right artery, it occurs in an isolated form, and is usually asymptomatic. Agenesis of the left pulmonary artery is associated with other cardiovascular malformations, such as tetralogy of Fallot, ventricular septal defect, aortic coarctation, pulmonary stenosis, and persistent arterial duct, which present more symptoms. When it is not associated with these types of disorders, it is considered an isolated finding.[3]

The most frequent mean age of diagnosis is around 14 years old. It is estimated that 13% to 30% of patients may remain asymptomatic for many years, and the disease is detected as a chance finding when performing a chest X-ray.[4]

Patients may present with exertional dyspnea, persistent cough, hemoptysis due to excessive aorto-pulmonary collateral circulation from either hypertrophied bronchial collateral vessels or ipsilateral peripheral arteriovenous fistulas to the absent pulmonary artery (20% of cases), chest pain, respiratory distress, heart failure, recurrent respiratory infections, or pulmonary hypertension in 25% to 44% of cases.[5]

The physical examination is normal. Cardiac murmurs and pulmonary hypoventilation can be auscultated, with or without pathological sounds in the affected hemithorax.[6]

Definitive diagnosis is based on imaging tests. The chest X-ray is the first line of investigation, showing a size-reduced lung in the agenesis hemithorax with ipsilateral mediastinal displacement, ipsilateral hemidiaphragm elevation, associated with contralateral pulmonary hyperinflation.[7] The diagnosis is confirmed with contrast-enhanced tomography. The parenchyma may present mosaic attenuation patterns, emphysematous changes, or bronchiectasis secondary to compensatory changes or recurrent infections.[8]

The echocardiogram can show associated cardiovascular malformations, pulmonary hypertension, and cardiac dextroposition. In most patients, collateral circulation develops from the descending aorta, especially the abdominal.[9]

The SPECT (single-photon emission computed tomography) shows exclusion of complete perfusion on the affected side, with normal ventilation. Cardiac catheterization is necessary when revascularization is planned.[10]

The multiphase MR (magnetic resonance) angiography allows for obtaining anatomical and functional information of thoracic vascular structures with a single injection of intravenous contrast and without the use of ionizing radiation.[11]

Lung function tests show normal or restrictive pattern with normal diffusion capacity.

The electrocardiogram is usually normal but may show right ventricular dominance in cases associated with pulmonary hypertension[12]

Conservative treatment is indicated for asymptomatic forms, and surgical treatment for cases with symptoms or serious complications.

Surgical management by lobectomy or pneumonectomy with selective embolization is performed in patients with massive hemoptysis or recurrent lung infections. Repair surgery consists of two stages: surgical anastomosis of the proximal and distal parts of the pulmonary artery, followed by *stent implantation*.[13]

Approximately 19% to 25% of patients with congenital absence of the pulmonary artery present with pulmonary hypertension later in life, which implies a poor prognosis, with a mortality rate of approximately 7%. This can be treated with antihypertensive drugs.

Death can be secondary to massive hemoptysis, respiratory failure, pulmonary hypertension leading to right heart failure.

No cases of pulmonary thromboembolism associated with unilateral absence of the pulmonary artery have been described; however, chronic thromboembolic pulmonary hypertension has an incidence of 0.01% to 9.1% after an acute pulmonary embolism and can be resolved by thromboendarterectomy.[14]

Volkan et al. reported a case of left pulmonary artery agenesis without congenital cardiac anomalies, similar to the patient described in this review, which could contribute to the maintenance of an asymptomatic state.

The diagnosis of unilateral agenesis of the pulmonary artery can be considered a challenge, and a high level of suspicion would help obtain a proper diagnosis.[15]

Conflicts of interest

The authors have no conflict of interest to declare.

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