Case 6/2008 – Thirteen-month-old Male Infant, with Systemic-Pulmonary Collateral Vessel into the Left Inferior Lobe, with Possible Arteriovenous Fistula in the Same Lobe, with a Structurally Normal Heart

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Clinical Data

A cardiomegaly was discovered during chest x-ray indicated due to pneumonia at 7 months of age. Slight fatigue was noticed since then and the patient was kept without medication.

Physical examination

The patient presented slight tachypnea, was acyanotic (98% capillary saturation) and pulses were normally palpated in the upper and in lower limbs. Weight was 12 kg, height= 78 cm, BP= 90/60 mmHg, HR= 128 bpm. The aorta was not palpated at the suprasternal notch.

The precordium showed no deformities or impulsions. The ictus cordis was palpated in the 4th left intercostal space in the hemiclavicular line and was limited by one digital pulp. The second heart sound was slightly hyperphonetic in the pulmonary area and a systolic ejection murmur was ausculted in the mitral area and the dorsum to the left.

The lungs were well ventilated and had no adventitious sounds; the liver was palpated at 1.5 cm from the right costal border.

The electrocardiogram showed a sinus rhythm and signs of left ventricular overload, considering the presence of accentuated QR waves of V4 to V6, with ÂQRS in +40º, ÂP:+60º and ÂT: +10º.

Radiographic imaging

The radiographic imaging showed slightly augmented cardiac area, with elongated and rounded left ventricular arch and normal pulmonary vascular network. The middle arch was concave. It was noteworthy a retrocardiac hypertransparent image in the left lower lobe, which shows the more prominent pulmonary vascular network with indistinct borders (Figure 1).

Key Words

Arteriovenous fistula; cardiomegaly; mitral valve insufficiency.

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Diagnostic impression

This radiographic image is compatible with the diagnosis of possible accentuated arteriovenous pulmonary fistulae in the lower left lobe, or systemic-pulmonary vessels, with a consequent left ventricular increase.

Differential diagnosis

An increased left ventricle with normal pulmonary vascular network could also suggest a cardiopathy with mitral valve failure or even dilated myocardiopathy.

Diagnostic confirmation

The clinical elements were compatible with the initial diagnosis of mitral failure, corroborated by the echocardiogram, which showed a slight increase of the left cavities. However, there were no signs of regurgitation at the level of mitral valve, but a systemic-pulmonary flow in the descending aorta due to possible collateral vessel. An angiotomography showed a large-caliber collateral vessel with 7 mm of diameter in the inferior descending aorta that continued with the left lower lobar artery. From then on, the large-caliber and winding vessels in the left lower lobe were visualized, with rapid drainage through the inferior pulmonary vein to the left atrium (Figure 2). The cardiac catheterism showed this same angiographic aspect, with a rapid pulmonary venous return flow to the left cavities (Figure 2). The lung scintigraphy with technetium 99m showed the lack of mapping in the left lower lobe, with a right pulmonary flow of 60%, left pulmonary flow of 30.2% and systemic flow of 9.7%. A selective injection of the radionuclide directly into the systemic-pulmonary collateral vessels showed that 30% of this flow shunted to the systemic side, possibly characterizing the presence of an arteriovenous fistula in this lobe.

Procedure

Considering the disconnection with the pulmonary arterial tree, in addition to the pulmonary arteriovenous fistula, a left inferior pulmonary lobectomy was performed, after the dissection and ligature of the systemic pulmonary vessel, the left inferior lobar artery and the left inferior bronchus. A pulmonary biopsy of this lobe showed Heath-Edwards grade IV pulmonary arterial hypertension. The patient's evolution was good.

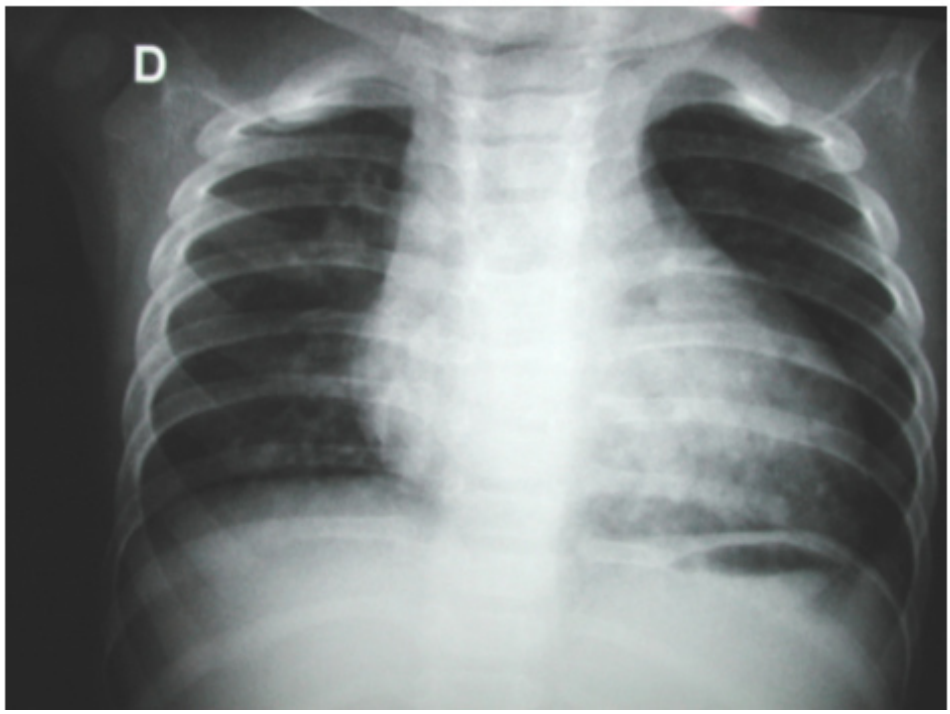

Figure 1 - Chest x-ray showing slightly increased cardiac area, with elongated and rounded left ventricular arch; The pulmonary vascular net is normal; The retrocardiac condensation image to the left in the lower lobe is clear, compatible with arteriovenous fistula.

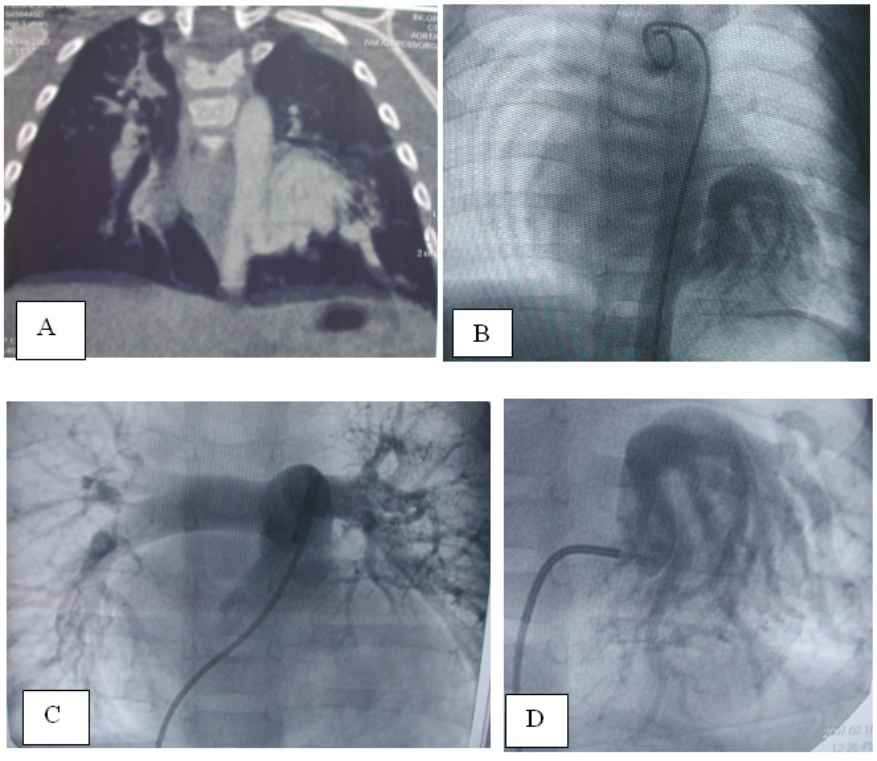

Figure 2 - Images with contrast show the presence of a systemic-pulmonary collateral vessel arising from the descending aorta in A (angiotomography) and in B (arterial angiography), with twisted and high-caliber arteriovenous vessels, exteriorized mainly in D; The injection in the pulmonary artery shows the absence of left lower lobe filling, in C.

Considerations

Without demonstrated cardiopathies, but with arteriovenous pulmonary fistulae, the absence of cyanosis can be explained by an arteriovenous blood shunt, already saturated from the aorta to the left inferior lobe, which suggests, at a final analysis, to a type of pulmonary sequestration.

Given the magnitude of this blood shunting, the increase in the left ventricle in the chest x-ray and the left ventricular overload at the electrocardiogram can be explained (Figure 2). The pulmonary lobectomy was deemed to be more effective and lower-risk than the connection of the left inferior lobar artery with the rest of the pulmonary arterial tree, especially considering the existing accentuated and irreversible degree of pulmonary hypertension.