CARDIOLOGIE TUNISIENNE

CASE REPORT

Unusual cause of heart failure in infants: Pulmonary sequestration : case report and literature review

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Summary

Introduction: Pulmonary sequestration (PS) is the second most common congenital lung malformation defined by a non-functioning lung tissue separated from the normal bronchopulmonary tree and vascularized by an aberrant systemic artery. The clinical presentation is heterogeneous and may rarely be revealed by congestive heart failure in infants.

Observation: We report the case of an 8-month-old infant hospitalized for persistent respiratory distress and signs of heart failure. Echocardiogram showed unexplained left heart dilation without evidence of an intracardiac shunt. The diagnosis was confirmed by CT angiography showing intralobar pulmonary sequestration.

Conclusion:Left to right shunts are the main cause of heart failure during the first year of life. Pulmonary sequestration is a rare cause of heart failure that should not be dismissed and should be considered in the presence of cavity dilatation without evident shunt.

Keywords

Pulmonary sequestration, heart failure

Résumé

Mots-clés

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INTRODUCTION

Pulmonary sequestration (PS) is a rare congenital lung malformation, with an estimated incidence of 0.15–1.8%, and constitutes the second most common congenital lung malformation (1). It is defined as a non-functioning lung tissue separated from the normal bronchopulmonary tree and vascularised by an aberrant systemic artery (2). We usually define two types of PS: intralobar (75% of cases) and extralobar (25%), depending on whether or not the abnormal lung tissue possesses its own pleural covering. The abnormal venous drainage of the sequestered lung could occur through the pulmonary vein into the left atrium, thus, creating a unique "left to-left" shunt and, as a consequence, left cardiac chamber enlargement. Less commonly, the venous drainage occurs into systemic circulation through intercostal veins, azygous–hemiazygous system, superior vena cava, or inferior vena cava, creating a left-to-right shunt and dilated right chambers (3). We report here a case of intralobar sequestration revealed by congestive heart failure in an 8month-old infant with spontaneous involution of pulmonary sequestration

OBSERVATION

An 8-month-old boy was referred to pediatric emergency for fever, tachypnea, persistent respiratory distress, and poor feeding. The clinical assessment revealed a continuous murmur, tachycardia (150 ppm), tachypnea (65 bpm), and hepatomegaly. Chest x-ray showed cardiomegaly with abnormal opacity in the lower left lobe, diagnosed at the time as pneumonia. The echocardiogram showed unexplained left heart dilation without evidence of an intracardiac shunt (Figure n°1). The ventricular function was normal. However, there was unusual color turbulence at the left inferior pulmonary vein (figure n°2). Left cavities dilatation with the Doppler pattern in the pulmonary vein raised the suspicion of pulmonary sequestration. Then, we performed CT angiography, which showed lobulated intra parenchymal-mediobasal left retrocardiac formation with vascular enhancement without parenchymal portion nourished by systemic artery arising from the left anterolateral side of the descending aorta with venous drainage to the left inferior pulmonary vein (Figure n°3). Mosaic perfusion of the left lower lobe was also noticed. The final diagnosis was intralobar lung sequestration with anomalous blood supply from the descending thoracic aorta and venous drainage via the pulmonary vein. Diuretic treatment was administered with a slight improvement in respiratory signs. We decided to postpone the surgery because of the improvement under medical treatment.

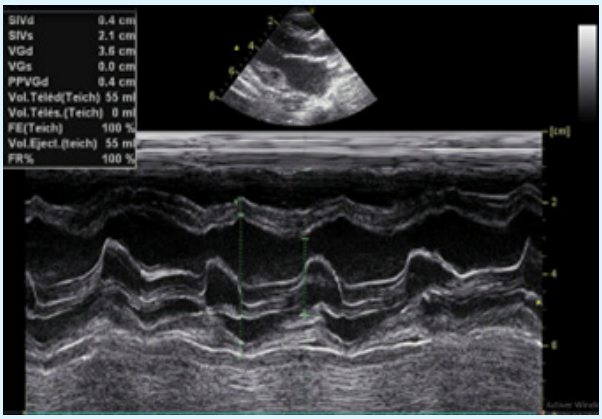

Figure 1. left ventricular dilatation

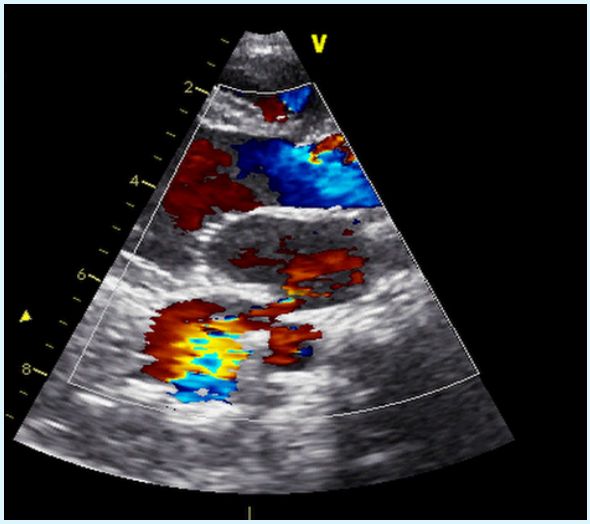

Figure 2. parasternal view: color turbulence at the left inferior pulmonary vein

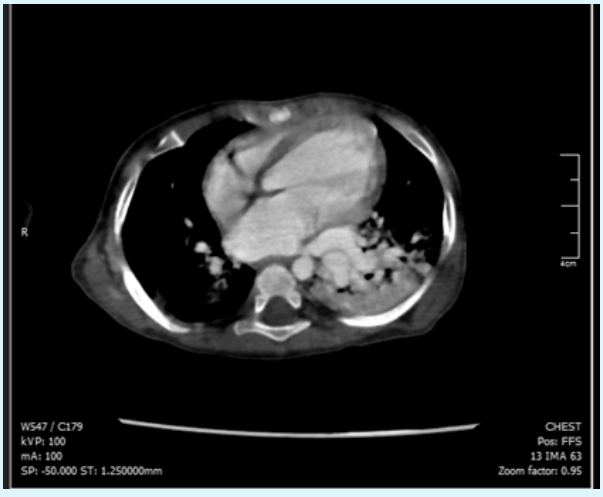

Figure 23 CT scan showing venous drainage of the systemic artery to the left inferior pulmonary vein

DISCUSSION

The original description was made by Pryce in 1946, who described three types of intralobar abnormalities based on the distribution of the aberrant systemic artery. In type 1, the anomalous artery supplies functional lung tissue, which communicates with the tracheobronchial tree. In type 2, the artery supplies both normal lung tissue, as well as non-functional, non-communicating tissue. In type 3, the anomalous artery supplies the lung, which is isolated from the tracheobronchial tree (4). Many variants are actually recognized not adhering strictly to the original definition and two principal types are described: intralobar and extralobar sequestration.

In intralobar sequestration (ILS), the abnormal parenchyma is included in the

parenchyma of the normal lung and is enveloped by the same visceral pleura; the usual site is the posterobasal region of the lower lobe without predominance of side. The arterial supply is derived from the thoracic aorta and venous drainage occurs through the pulmonary veins with the resultant left-to-left shunt. They usually have a late presentation with respiratory distress and difficulty in feeding. Some cases of severe, even life-threatening hemoptysis, have been reported (5). Rarely do they present with evidence of congestive heart failure secondary to excessive blood flow through the sequestered lobe (6).

Extralobar sequestrations (ELSs) do not have a connection with the normal lung due to their development from a supernumerary accessory tissue. There is, therefore, a complete anatomical and physiological separation from the normal parenchyma. ELSs have their own independent visceral pleura. In 75% of cases, they are located between the diaphragm and the lower lobe (80% on the left). The other locations in the middle and upper lobes are rare. The venous drainage is usually via the inferior caval vein, portal venous system, or azygous vein, resulting in a left-to-right shunt. They are in presenting early in life and have typical manifestations of a left-to-right shunt lesion (7). This type of sequestration is often associated with other congenital anomalies (such as diaphragmatic hernia, bronchopulmonary foregut duplication cysts, and cardiac defects) and with CHD, including truncus arteriosus, total anomalous pulmonary venous return, pulmonary atresia with ventricular septal defect, and dextrocardia (8,9).

Our patient had typically mediobasal intralobar sequestration, supplied from the descending thoracic aorta.

The foremost objective of imaging, when bronchopulmonary sequestration is suspected, should be to show the aberrant vascular anatomy.

Doppler echocardiography allows suspecting the diagnosis by highlighting signs of the shunt with dilatation of the cardiac cavities without true communication. It is also ideally suited for evaluation in the antenatal period or in neonates. PS should be considered in the differential diagnosis of a fetus with a lung mass (10,11). Many extralobar pulmonary sequestrations decrease dramatically in size in utero and may not need treatment after birth (12).

Multidetector CT angiography is the current diagnostic imaging method of choice for optimum evaluation of the sequestered lung and its vascular supply. Computed tomography can also help in diagnosing other lung conditions that may mimic sequestration on a chest radiograph. Multiplanar CT reconstructions and 3-D reconstructions are useful in depicting the arterial and venous anatomies of a sequestered lung segment (13,14, 15). MRI is an alternative imaging method providing excellent imaging of the supplying systemic arterial anatomy (16).

Treatment of pulmonary sequestration is necessary due to the risk of recurrent respiratory infection and spontaneous hemothorax.

Surgical resection remains the treatment of choice for pulmonary sequestration. Classically, sequestered lung segments have been resected via thoracoscopic surgery or thoracotomy. Transcatheter embolization of systemic arterial supply of sequestered lung segments using embolization coils and devices has also been reported (17,18). Several cases of spontaneous involution have been reported in the literature (19).

CONCLUSION

Left to right shunts are the main cause of heart failure during the first year of life. Pulmonary sequestration is a rare cause of heart failure that should not be dismissed and should be considered in the presence of cavity dilatation without evident shunt.

The limits of this study are the retrospective type and the small size of the studied population. A prospective and multicentric study must be conducted to evaluate the results of this approach.

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