Pulmonary Hypertension in Infants Linked to Horseshoe Lung: Case Report

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This is the case report of a young infant with early respiratory distress and pulmonary hypertension, diagnosed as a variant of horseshoe lung and we have reviewed the literature to seek information about this rare pulmonary malformation and its cardiac and hemodynamic repercussions.

Introduction

Horseshoe lung is a rare congenital anomaly defined as a fusion of the lower posterior segments of the lungs. The involvement of the right lung with hypoplasia is more common and in most cases, it is associated with the scimitar syndrome. The involvement of the left lung is exceptional and related to worse outcomes, as well as the presence of associated cardiac malformations and pulmonary hypertension[1,2].

We report the case of a young infant referred to pediatric cardiology center due to cyanotic episodes.

Case Report

This article reports the case of an infant at three months of life, female, referred to a referral center of pediatric cardiology for presenting dyspnea since birth and frequent episodes of cyanosis. After birth, the patient developed respiratory distress and mild hypoxia in the early hours of life, requiring support with mechanical ventilation. The patient remained in the neonatal intensive care unit (ICU) for 30 days and was discharged after 40 days of life still with respiratory distress and feeding difficulty. At two months of life, she had an episode of cyanosis, pneumonia was diagnosed on the left, requiring ventilatory support for 04 days and specific antibiotic therapy. As there was no improvement after therapy, persisting with frequent episodes of cyanosis, she was referred to a tertiary pediatric cardiology service.

Keywords

Heart defects, congenital; hypertension, pulmonary; infant

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Upon admission, she had an impaired general health status, was moderately tachydyspneic, with mild central cyanosis using a nasal oxygen catheter (O_2) 3 l/min. Upon inspection of the chest, the patient had precordial bulge and hyperdynamism and intense respiratory distress. Upon cardiac auscultation, the patient was in regular cardiac rhythm, tachycardic, with accentuation of the pulmonic component of the second heart sound, without murmurs. Upon lung auscultation, there was evidence of decreased vesicular murmur on the left and diffuse subcrepitant rales, and mild inspiratory stridor. Pulses were present and symmetrical. In the abdomen, liver was palpable at 4.0 cm from the right costal edge.

The patient was admitted to the cardiopediatric ICU for hemodynamic stabilization where noninvasive ventilation was initiated with a fraction of O_2 of 0.5, furosemide 3.0 mg/kg/day and dobutamine, 5.0 μg/kg/min. Evolved with clinical stabilization, weaning from inotropic and ventilatory support, so that in the fifth day after admission, she was in ambient air with mild tachydyspnea, keeping O_2 saturation around 96.0% using furosemide 3.0 mg/kg/day and digoxin 0.01 mg/kg/day. Clinical research was then initiated.

Chest radiography showed a reduced volume of the left lung and deviation of mediastinal structures to this side (Figure 1a). The patient had, from the source hospital, computed tomography, whose local evaluation diagnosed hypoplastic left lung. Subsequent analysis, despite the poor technical quality of the test, also detected an image suggesting continuity between the lower lung lobes (Figure 1b).

Transthoracic echocardiography identified an increase of right heart chambers with signs of pulmonary arterial hypertension and small ostium secundum atrial septal defect (ASD).

Cardiac catheterization revealed significant pulmonary hypertension (Table 1).

On angiography, we observed right pulmonary artery branch going to the isthmus of the left lung, and is did not clearly demonstrated the origin of the left pulmonary artery (hypoplastic left lung), with venous return parallel to this pulmonary artery branch to the pulmonary isthmus draining into the left atrium (Figures 1c, 1d and 1e).

After cardiac catheterization, the child was readmitted to the ICU for recovery room and began drug therapy for pulmonary hypertension with sildenafil at the doses recommended. On the third day after catheterization, she was in ambient air, with the previously prescribed medications, awaiting discharge to the ward, when she had a sudden onset of cyanosis with sinus bradycardia progressing to asystole unresponsive to resuscitation, progressing to

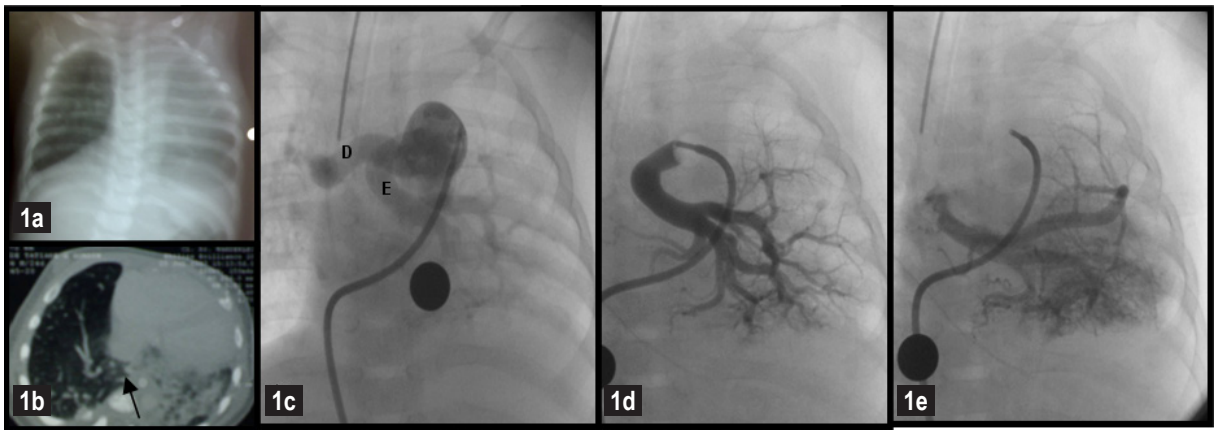

Figure 1 - 1a: Chest radiography in posteroanterior projection (PA) - shows deviation of structures from the mediastinum to the left hemithorax; 1b: Thorax tomography - pulmonary parenchyma in the thoracic midline (isthmus-arrow) and hypoplastic left lung; 1c: Angiography of pulmonary trunk in PA showing angiography of the pulmonary trunk in PA showing the right pulmonary artery (D) and its branch leading to the isthmus to the left lung (E); 1d: Selective angiography in this branch to the left lung - aberrant course and branches to the midline; 1e: Pulmonary isthmus venous return, which is parallel to the arterial branch, which drains into the left atrium.

Table 1 – **Pressure and oxygen saturation in the chambers specified and hemodynamic calculations.**

Cavity	Systolic pressure (mmHg)	Diastolic pressure (mmHg)	Average pressure (mmHg)	SO_2 (%)
SVC			11	60
PT	94	54	73	65
LA			12	99
AO	80	43	56	97
	Qp:Qs	RVP (UWxm²)	RVS (UWxm²)	RVP/RVS
	1.09	18.80	15.10	1.246

SO_2 - Oxygen saturation; SVC - superior vena cava; PT - pulmonary trunk; LA - left atrium; AO - aorta; Qp:Qs - systemic and pulmonary flow ratio; PVR - pulmonary vascular resistance; SVR - systemic vascular resistance; UWxm² - Wood units per square meter of body surface.

death. The episode was interpreted by an assistant on duty as "pulmonary hypertension crisis".

Although aware of the severity of the condition, and informed about the importance of laboratory diagnosis, the mother did not authorize anatomopathological study, which made it impossible to confirm the anatomy of the data reported here.

Literature review and considerations

Horseshoe lung is characterized by the presence of the isthmus of pulmonary parenchyma that extends across the midline, connecting the posterior basal regions of both lungs, behind the heart and anterior to the descending aorta. Since its description in the 60s (Spencer, 1962) about 40 cases were reported in the world literature, mostly associated with hypoplastic right lung and often associated with scimitar syndrome. There is only one case described in a fetus of 19 weeks, where the pulmonary isthmus was located in the apical portion of the lungs, which led to reflections about the embryological origin of this malformation[1-3].

In most cases, the clinical symptoms began early with lung problems in young children, infants and neonates. Frequent respiratory distress, recurrent pneumonia and episodes of cyanosis are the most common findings. Pulmonary hypertension has been implicated in the early evolution of such patients, although it is not a rule[1,4]. The scimitar syndrome is associated in 15.0% of the cases with horseshoe lung. Despite the morphological similarities between the syndrome with or without this association, its clinical presentation is different[5]. Patients with horseshoe lung have obvious symptoms early and early need for surgery, as discussed below. Congenital heart defects occur in 25.0% of the cases; the most common one is the atrial septal defect (50.0%). Other lesions include ventricular septal defect, patent ductus arteriosus, tetralogy of Fallot, total anomalous pulmonary veins, in a case described recently in the national literature, and coarctation of the aorta[6,7].

Our patient presented symptoms very early in life, with severe pulmonary hypertension, associated atrial septal defect and anomalous pulmonary venous drainage, factors related to poor prognosis.

In chest radiography there is hypoplasia of one lung and deviation of mediastinal structures to the same side, but no typical finding of malformation[1]. Chest computed tomography demonstrates unilateral pulmonary hypoplasia with isthmus extending beyond the midline. Generally, the right lung is affected. However, as in the case reported here, cases of involvement of the left lung have been described. Also, it has been observed that the isthmus originates from the hypoplastic lung in all cases[2,8]. The definition of the diagnosis is made by angiography or bronchography when the bronchial and anomalous vascular supply to the pulmonary isthmus is defined. Cardiac catheterization, in addition to demonstrating the arterial vascular anatomy, is an important tool in the evaluation of venous drainage of the affected lung, and there may be or not alteration and obstruction in its path, and in the diagnosis and hemodynamic study of cases associated with pulmonary hypertension[1,2].

In our case, the diagnostic possibility was suggested only after cardiac catheterization, where treatment with Sildenafil was instituted in an attempt to minimize symptoms related to pulmonary hypertension, which appears to have greatly contributed to the outcome of the case.

When possible, surgical treatment is indicated in the presence of recurrent infections, significant left-right short-circuiting (Qp:Qs > 2:1) in the presence of associated cardiac defects and progressive pulmonary hypertension. The correction of existing intracardiac anomalies is strongly recommended, or if there is recurrent infection, resection of the affected lung. Due to the poor prognosis of these patients postoperatively, if there are no symptoms or significant systemic-pulmonary short-circuiting, conservative treatment is recommended[1].

Here, we report a rare case of horseshoe lung with involvement of the left lung, stormy clinical course and severe pulmonary hypertension. Since it was not possible to perform an anatomopathological study of the case, some comments are needed. Though described in isolation as presented here, horseshoe lung is commonly related to the scimitar syndrome. Thus, in isolation, the aberrant origin of the left pulmonary artery could characterize a form of vascular ring, such as the sling of the left pulmonary artery, but without compression of the distal trachea. We could also question whether the abnormal course of this artery would not be a variation of the pulmonary arterial branch.

Anyway, we consider important to describe the case so that the recognition of this disease be possible in its early stage, as it presents an association with variable degrees of pulmonary hypertension of an evolutive character and, in general, poor prognosis.

Potential Conflict of Interest

No potential conflict of interest relevant to this article was reported.

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Study Association

This study is not associated with any post-graduation program.

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