Obstructed right pulmonary venous drainage and ipsilateral lung hypoplasia

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Abstract

A case of right lung hypoplasia and dyspnea, presented at our institution for cardiac evaluation, is described. Clinical evaluation and instrumental findings of normally connected but completely obstructed right pulmonary venous drainage have led us to diagnose an exceptional variant of subtotal cor triatriatum. Differential diagnosis was mainly carried out between pulmonary venous anomalies and drainage obstructions. Symptom relief was obtained with surgical excision of the atrial dividing membrane.

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1. Introduction

Congenital pulmonary venous disorders comprise a wide range of clinical presentations and associated lesions [1]. Features are not distinctive and only a strong index of suspicion facilitates instrumental appreciation. Lung hypoplasia (LH) is usually associated with multiple congenital malformation syndromes, but unilateral involvement is often linked to structural heart disease.

Herein we describe a case earlier categorized as isolated right LH; subsequently normally connected but completely obstructed pulmonary venous drainage was discovered and treated.

2. Case description

The patient, a 10-year-old girl with effort-related dyspnea, was referred at our institution for cardiac evaluation. She was previously diagnosed with congenital right LH, otherwise her medical history was unremarkable. On physical examination (weight 35 kg, height 135 cm) the heart sounds were almost normal and no murmurs were appreciated. Chest roentgenogram was notable for a smaller right hemithorax, associated to mild mediastinal shift, and for ipsilateral vascular congestion.

Echocardiographic study revealed a normal-sized left atrium (LA) connected to dilated left pulmonary veins (PVs). The right ones were not visualized. Search for anomalous connected veins and atrial septal defect was negative. The other findings were mild right ventricular dilatation and pulmonary arteries asymmetry (right 15 mm; left 8 mm). Doppler examination suggested bi-directional flow in the right pulmonary artery. At catheterization, systemic saturation was 99% and no oximetry step-up was found, but pathological pressure was recorded in the right pulmonary artery (30/13/19 mmHg, wedge 29 mmHg) (left wedge 9 mmHg). Finally, angiography showed an hypoplastic right pulmonary tree and an anomalous right venous drainage into a closed formation bulging into the LA (Figs. 1 and 2). All right venous return was collected in this cul-de-sac and retrograde catheterization was not possible. No anomalous systemic arterial supply was found.

The patient underwent elective surgery to relieve the obstructed pulmonary right drainage. Median sternotomy was performed and the pulmonary veins were fully exposed: right LA connection was normally apparent and no stenotic or ectasic vein was visualized. Operation was continued on cardiopulmonary bypass/cardioplegic arrest and with moderate hypothermia. Transverse right atriotomy was made and the blind chamber was entered prolonging the incision through the septum at fossa ovalis level.
A membrane dividing the cul-de-sac and the LA was visualized and the PVs were normally apparent without stenosis. The membrane was fully excised. Glutaraldehyde-fixated autologous pericardium was then employed for augmentation of venoatrial junction and septum closure. At careful inspection, a pinhole meatus was discovered in the middle of the membrane. Postoperative course was uneventful. The patient reported symptom relief and echocardiogram was almost normal at early follow-up.

3. Discussion

The primitive splanchnic plexus drains venous blood from the lung buds into umbilical vitelline and cardinal veins. The common pulmonary vein grows out as an evagination from the LA side of the common atrium and joins pulmonary veins while systemic venous connections disappear. As the left atrium grows, common pulmonary vein is incorporated into its posterior wall and PVs enter the LA separately. Derangement of this process at different stages results in a broad spectrum of anomalies: partially or totally anomalous pulmonary venous connection, common pulmonary vein atresia (CPVA), cor triatriatum sinister, stenosis or abnormal number of PVs [1].

The reported clinical and instrumental findings were interpreted mainly according to the following considerations.

PV stenosis is a morphologic and functional narrowing of the junctional area between one or more PVs and the LA. It may be categorized into localized stenosis or diffuse hypoplasia (even atresia) [2]. It may be further subdivided in congenital or acquired (secondary to mediastinal fibrosis or complicating surgery for anomalous venous connection, transplantation, arrhythmias) and the prognosis is usually poor. The outcome is surely related to the number of involved vessels and associated cardiac defects and progression to pulmonary vascular disease is usually reported [3–5]. Some clinical and instrumental findings could suppose unilateral slowly progressive stenosis for this case, but standard obstruction levels do not fit well with the morphologic features observed [2,6].

CPVA provides for normally formed and connected veins to a chamber that ends blindly behind the LA, without connection between either left or right atrium or a systemic vein. The cul-de-sac appearance at right veins’ confluence strongly resembles the blind chamber required by the definition of CPVA, but clinical context (different from postnatal cardiac and respiratory failure) and unilaterality of the lesion do not allow this denomination for the case [7,8].

Cor triatriatum results from derangement of total incorporation of the common pulmonary vein into LA. The pathological finding is a membrane that divides the LA into two chambers: a proximal one that receives normally formed PVs and a distal one that includes appendage and mitral vestibule [7,9]. The dividing membrane usually contains one or more openings. The proximal chamber may communicate with the right atrium either directly or via an anomalous venous channel. If the proximal chamber does not receive all PVs, subtotal cor triatriatum is defined [7,9]. The natural history of this rare condition has been poorly documented.

Finally, multiple malformation syndromes may be found in infants with LH. Particularly unilateral right involvement, in the absence of an intrathoracic mass, is strongly
associated with cardiac anomalies [10]. In the reported case the right lung was uniformly affected and no findings of partially anomalous pulmonary connection or systemic arterial supply were present.

The clinical picture and the literature review have allowed us to define the described obstructed right pulmonary venous drainage as a rare case of subtotal cor triatriatum. So far as we known, it is an unreported variant of the type C1a, according to Herlong et al. [7] or A2, according to Thilenius et al. [9], since the proximal chamber was not provided with effective outflow channels. Ipsilateral LH may be considered a secondary or synchronous disorder to this kind of pulmonary venous derangement.

In conclusion, we have described the process of differential diagnosis that led to the identification and treatment of an extreme form of left atrium partitioning. Division of left atrial chamber and the group of pulmonary venous anomalies deserve particular attention to promptly address surgical correction.

References