Case Report

A Rare Association of a Cor Triatriatum Sinistrum and a Right Partial Anomalous Pulmonary Venous Return: A Route to Understanding the Disease

Tueche, S.
Department of Cardiac Surgery of Brugmann, University Hospital. Place A Van Gehuchten 4, 1020, Brussels, Belgique.

Abstract

A 56 day-old baby girl referred for recurrent respiratory tract infections and stridor, showed a rare association of a Cor Triatriatum Sinister (CTS) and a right Partial Anomalous Pulmonary Venous Return (PAPVR), in the form of a stenosed Scimitar vein, draining the right pulmonary veins to the subdiaphragmatic inferior vena cava (IVC), on echography and angiography. The baby underwent successful surgical repair of the CTS, by resection of the obstructive membrane and reconnection of the Scimitar vein to the left atrium. At the age of 5 years, the child is doing well without any evidence of obstruction on the right or the left pulmonary venous drainage.

Key words: Partial Anomalous Pulmonary Venous Return (PAPVR), Cor Triatriatum Sinistrum (CTS), Scimitar vein.

Case Report

A 56 day old baby girl was admitted for repeated pulmonary infections and failure to thrive. Clinically, there was a marked stridor and tachypnea. The mental development was normal and there was no clue for an abnormal phenotype. No family history of congenital heart disease was noted. A detailed workup, including conventional chest roentgenography, echography, bronchoscopy and angiography led to the diagnosis of left bronchial compression by a distended left atrium, due to a CTS (Fig-1). As the infant failed to respond favourably on maximal medical support, urgent surgery was decided. Through a median sternotomy, on deep hypothermic (20°) cadiopulmonary bypass the left atrium was exposed through the right atrium and the atrial septal defect. There was a stenosing perforated membrane between the hypotrophic left atrial cavity and the retroatrial pulmonary venous chamber collecting only the left pulmonary veins. The abnormal membrane was resected and the connection of the left pulmonary venous chamber with the left atrium augmented with a patch of autologous pericardium. The right pulmonary venous drainage could not be located and the operation was concluded for fear of prolonging excessively the aortic cross clamp time. The child recovered well with some improvement of the respiratory status but persistent stridor and tachypnea. A second angiography was performed, which diagnosed an anomalous pulmonary venous drainage of the right lung to the IVC, in the form of a typical Scimitar vein (Fig-2), which was stenosed at the subdiaphragmatic IVC junction. There were also a few systemic feeding arteries from the abdominal aorta to the right lower lung (Pulmonary sequestration). The infant was taken again to the operating room and the chest was reopened. The Scimitar vein was easily identified after opening the right pleura. On cardiopulmonary bypass with a 10 minutes period of deep hypothermic total circulatory arrest the vein was disconnected from the IVC and anastomosed to the right lateral wall of

Correspondence: Serge Tueche, General Surgeon and Clinical Scientist, 17 Rue du Gros Chene, Boite 207, 44300 Nantes, France. Tel: 0033.665.220.155.
E-mail: serge.tueche@wanadoo.fr

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the left atrium. The systemic arteries to the right lower lobe were ligated. The infant was extubated on the second postoperative day, recovered uneventfully and is still doing well 5 years later at follow-up, with a normal growth curve and no echographic suggestion of stenosis on the right and left pulmonary venous drainage.

Discussion

This case is unusual because of the coexistence of a CTS and a right PAPVR. CTS is among the rarest malformations comprising less than 0.1% of all antemortem diagnosed congenital heart disease and 0.4% of necropsy series, with a male predominance of 1.5/1[1-4], while, PAPVR is an anomaly seen in 1 % of patients with congenital heart disease, with no gender prevalence[3-5]. CTS was described the first time by Church in 1868[1], while the malformation of the membrane dividing the right atrium into an anterior and posterior chamber was first mentioned by Lauenstein[2]. The PAPVR in this case resembles a Scimitar syndrome. Despite the fact that our patient displayed angiographic characteristics of the Scimitar vein and typical pulmonary sequestrations of the right lower lobe, it was atypical, because the right lung was not

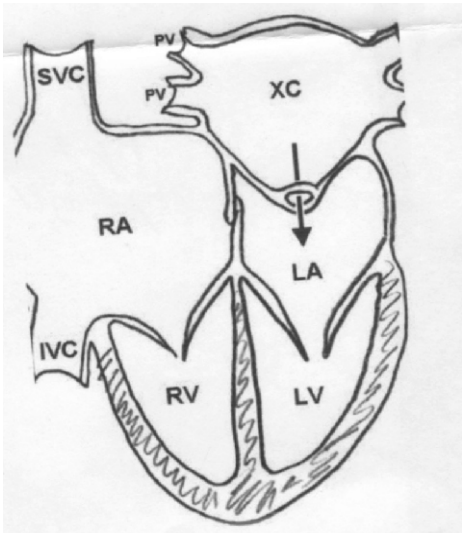


Fig. 1: Representation of CTS (arrow). Legend: SVC: superior vena cava. RA: right atrium. IVC. RV: right ventricle. PV: pulmonary veins. XC: collecting chamber. LA: left atrium. LV: left ventricle.

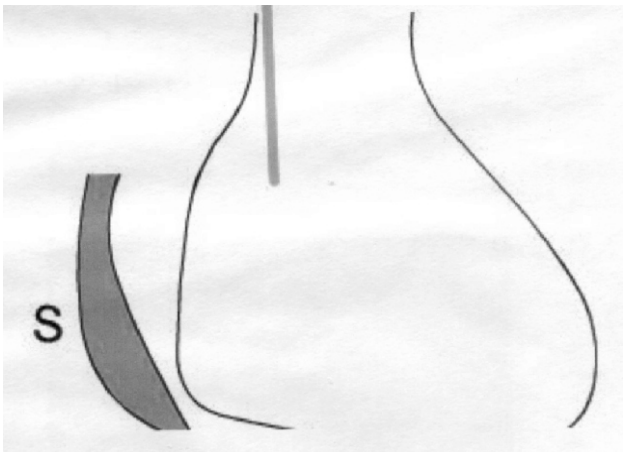


Fig. 2: Representation of the Scimitar vein.

hypoplastic and the right pulmonary arteries were normal in size and distribution. Embryologically, in CTS, the left atrium is divided in two distinct chambers, proximal and distal[1-4]. The proximal, or dorsal (posteroinfer) chamber corresponds to the common pulmonary vein which has failed to incorporate normally into the left atrium, at 35th days postconception. The distal, or ventral (anterosuperior) chamber, represents the true left atrium. Both chambers are separated by a variably perforated membrane, composed by the common pulmonary vein dorsally and the wall of the primitive left atrium ventrally[1-4]. In PAPVR, partial failure of the developing pulmonary venous plexus to join the left atrium with persistent connection to the primitive systemic venous system is the suggested explanation[1-5]. Anatomic studies show the right atrium to be enlarged and thick-walled in PAPVR. The left atrium is small (< 50 % of predicted normal), while the left atrial appendage is normal in size. The precarious clinical status of our patient was due, to the presence of severely obstructed CTS, together with an obstructed Scimitar vein. Echocardiography is the best diagnostic mean[3]. The Scimitar vein may sometimes be obvious on plain RX of the thorax. The goal of surgery in PAPVR is to achieve a patent anastomosis between the abnormal, partial pulmonary venous return and the left atrium and in CTS to complete excision of the

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false membrane[4]. In PAPVR, late pulmonary vein obstruction due to stenosis in the anastomotic area occurs in to 15% (mostly 6 to 12 months after surgery) due to diffuse fibrous hyperplasia originating from the atrial wall and invading the pulmonary venous ostia[5]. A lesson learned from this unusual case is that, if one does not find pulmonary veins within the pericardium, they should be examined by opening the pleura, in search for an extracardiac connection. This case also supports the hypothesis that PAPVR and CTS are closely related entities sharing a common developmental fault of the incorporation of the pulmonary plexus in the left atrium.

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