Case report - Congenital

Scimitar syndrome in an adult: diagnosis and surgical treatment

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Abstract

A 51-year-old woman, presenting with increasing dyspnoea on exertion was admitted for surgery of a right sided partial anomalous pulmonary venous return. Contrast enhanced electron-beam tomography (EBT) presented a hypoplastic right lower lobe and confirmed the diagnosis of a so-called scimitar syndrome. Surgery consisted of creation of an atrial septal defect, transposition of the anomalous vein and reocclusion of the artificial septal defect with a Dacron patch. One year after surgery, the patient’s physical strength increased to a normal level without dyspnoea, while three-dimensional imaging reconstructed from EBT demonstrated an intact operative situs.

Keywords: Scimitar syndrome; Three-dimensional computed tomography; Adult congenital surgery; Partial anomalous pulmonary venous return

1. Introduction

Partial anomalous pulmonary venous return (PAPVR) is a relatively uncommon congenital anomaly accounting for 0.5–1% of congenital heart disease. The characteristic abnormality is PAPVR of part or all of the right lung to the inferior vena cava (IVC), either below the diaphragm or at the junction of the IVC and the right atrium (RA). Drainage into the hepatic vein, the portal vein or the left atrium can occur as an isolated lesion or together with other cardiac abnormalities like an atrial septal defect (ASD)\cite{1}. The rare combination (3–5%) of an association of PAPVR, hypoplasia of the right lung and dextroposition of the heart is designated scimitar syndrome\cite{2}. The ‘scimitar’ sign is a characteristic chest roentgenographic finding of a crescent-like shadow in the right lower lung field that is produced by the anomalous venous channel. A left-to-right shunt is established because the anomalous pulmonary vein drains blood from the right lung into the IVC resulting in an increased risk of developing right ventricular failure due to long-standing right ventricular volume overload.

The diagnosis can frequently be made on a chest radiograph and heart ultrasound. Beside invasive measures like catheterization, sophisticated imaging techniques allow to make precise diagnoses. Three-dimensional (3D) reconstructions give the surgeon a clear picture of the malformation, in order to develop the appropriate operative strategy. To establish the diagnosis in our patient, in addition to routine preoperative examinations non-invasive, 3D imaging reconstructed from contrast enhanced electron-beam tomography (EBT) was performed.

2. Case report

A 51-year-old woman was admitted to the hospital because of increasing dyspnoea on exertion since 2 years, otherwise the patient was asymptomatic. Chest radiograph presented an irregular shadow in the right lower lung field and PAPVR was suspected. Cardiac catheterization showed right-sided PAPVR, which drained as a common trunk into the RA at the inferior cavo-atrial junction. Oximetry during right heart catheterization revealed a left-to-right shunt ratio of 59% ($Q_p:Q_s = 2.6$). EBT presented a hypoplastic right lower lobe and confirmed the diagnosis of a scimitar syndrome (Fig. 1) and the indication for surgery. After median sternotomy and initiation of total cardiopulmonary bypass (CPB), an ASD was created and the anomalous vein was transposed to reach the RA. The artificial defect was then covered with a Dacron patch resulting in total correction of the vitium, and the IVC was closed with
The postoperative course was uneventful. Nine months later, the patient’s physical strength was reestablished and EBT and 3D imaging demonstrated the complete correction of the heart (Fig. 2).

3. Discussion

Standard diagnostic measures for a suspected PAPVR consist of transthoracic and transesophageal echocardiography as well as invasive heart catheterization including quantitative determination of the pulmonary to systemic blood flow ($Q_p/Q_s$) by oximetry. The accuracy of oximetry is impaired by anatomical/mechanical difficulties to selectively depict all anomalous vessels. In addition, oximetry is of limited value when the anomalous connection is to the IVC, because of both reduced flow through the right lung and the contribution of highly oxygenated blood from the renal veins. Therefore, the use of further imaging techniques like contrast enhanced cardio computed tomography (CT) or multi-slice CT is useful to confirm the diagnosis, identify the anomalous pulmonary vein and demonstrate other associated abnormalities [3]. In our patient, non-invasive EBT and 3D imaging reconstruction provided an excellent demonstration of the situs and allowed for precise planning of the operation.

Most frequently, patients are asymptomatic in the absence of associated abnormalities. The extent of symptoms and physiologic disturbances are dependent on the degree of shunting, the number of anomalous veins, any associated valvular abnormalities and the presence of any concomitant cardiac or pulmonary disease [4]. Surgical correction is recommended for symptomatic patients or asymptomatic patients with a pulmonary-to-systemic blood flow ratio exceeding 1.5–2 because of their higher likelihood of progression to pulmonary hypertension and right ventricular failure [5,6]. Both criteria were fulfilled in our patient. Directly connecting the scimitar vein to the left atrium, as described recently by Brown et al. [7] was not possible because of extreme dextroposition of the heart. CPB had to be used to create a neo-ASD in order to connect the transposed PA on the level of the intra-atrial septum to the RA. The neo-ASD was closed with a Dacron patch similar to the suggestion of Torres and Dietl [8]. An intratrastral tunnel was not necessary, thus avoiding complications like stenosis in the IVC or at the scimitar reimplantation site.

References