Atrial septal defect with drainage of the inferior vena cava into the left atrium

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Abstract

Atrial septal defect (ASD) with drainage of the inferior vena cava (IVC) into the left atrium (LA) is a rare congenital anomaly. Few cases have been reported in the literature. We present a 17-year-old female with an ASD and an anomalous drainage of the IVC into the LA leading to cyanosis since early childhood. Diagnosis was documented by computed tomography (CT) angiography and confirmed intra-operatively. The patient underwent successful surgical correction with an uneventful postoperative course.

Keywords: Atrial septal defect; Inferior vena cava

1. Introduction

Drainage of the inferior vena cava (IVC) into the left atrium (LA) is an uncommon congenital disorder, and is even less common when associated with an atrial septal defect (ASD). Only few cases have been reported in the literature [1–6]. Because of this, the natural course, clinical manifestations and diagnosis are not well established. Surgery is the only option for correcting such a disorder.

2. Case report

A 17-year-old female showed signs of cyanosis since 4 years of age. Unfortunately, she never had a diagnosis despite being evaluated by many physicians. She presented to us with increasing cyanosis that was aggravated by exercise and, lately, even with minimal physical activity.

Physical examination revealed a well-developed 17-year-old female. Blood pressure and heart rate were normal. Cardiac auscultation revealed an ejection systolic murmur at the left upper sternum and fixed split of second heart sound. She was cyanosed with clubbed fingers.

Her room air saturation was 85% by pulse oximetry. Her hemoglobin was 17.8 g l⁻¹ and hematocrit 50%. Chest roentgenogram showed evidence of a dilated right ventricle with clear lung fields. Electrocardiogram showed normal sinus rhythm, P pulmonale, right ventricle enlargement, and right axis deviation. Transthoracic (TTE) and transesophageal (TEE) echocardiography revealed a large, secundum ASD, located in the posteroinferior part of the septum, with no posterior rim. The right ventricle was dilated with left-to-right shunt. Because this does not explain the chronic cyanosis, other concomitant pathology needed to be ruled out. Cardiac catheterization and computed tomography (CT) angiography were performed. The patient was seen by the pulmonologist, and major pulmonary causes of cyanosis were ruled out. Pulmonary function tests were normal. Right- and left-heart catheterization findings are summarized in Table 1.

Angiogram showed no patent ductus arteriosus and no evidence of any arteriovenous fistulous connections within the lungs or between the left atrium and the pulmonary artery. The CT angiogram clearly demonstrated drainage of the IVC into the LA (Fig. 1A and B); hence, the diagnosis of an ASD associated with anomalous drainage of the IVC into the LA was made. The patient was referred for surgical correction.

The operation was conducted via a midline sternotomy. The ascending aorta and both vena cavae were cannulated directly, taking care of cannulating the IVC as low as possible. Cardiopulmonary bypass under moderate hypothermia...
3. Discussion

Many variations in the developmental pattern of the IVC have been described, but few of those anomalies are significant cause of congenital heart disease. The IVC drainage into the LA is a rare congenital anomaly, and is less commonly associated with an ASD. Most of the published articles are individual case reports [1–6]. It may occur in conjunction with other anomalies, such as anomalous pulmonary venous drainage and pulmonary arteriovenous fistulae. It was first described by Gardner in 1955 as an autopsy finding [7]. It is suggested that the abnormality is due to the persistence of the right valve of the sinus venosus [3,8]. Embryologically, the sinus venosus receives the cardinal, umbilical, and vitelline veins. It communicates with the primitive atrium via an orifice that has a right and left valves related to its right and left horns. Normally, the sinus venosus migrates to the right, and the left valve disappears. The left horn becomes the coronary sinus. The right valve usually decreases in size, becoming the crista terminalis, Eustachian, and Thebesian valves. If the right sinus venosus valve persists and fuses with the superior part of the secundum septum, the IVC will drain into the LA.

This entity is different from a low or inferior vena cava secundum ASD, where a prominent Eustachian valve can result in shunting of blood from the IVC to the LA. If the surgeon is not careful, this can be mistaken for the inferior ASD rim, and he may iatrogenically divert IVC blood to the LA upon ASD closure, causing cyanosis [9,10].

Establishing diagnosis can be difficult as illustrated from the fact that most reported cases in literature were diagnosed in adulthood [3]. Many imaging modalities can be used to establish diagnosis. Munet et al. [4] used perfusion pulmonary scintigraphy and contrast echocardiography, whereas magnetic resonance imaging (MRI) was used by Burri et al. [3] In our case, the TTE and TEE revealed the ASD but failed to demonstrate the anomalous IVC drainage into the LA. However, the presence of cyanosis made us suspect an associated disorder and, therefore, further diagnostic modalities were undertaken. The diagnosis was eventually established by CT contrast angiography (Fig. 1(A)). It thus appears that with the use of echocardiography, angiography, MRI, and CT, one should be able to firmly establish the diagnosis. Once this is confirmed, the patient should be referred for surgery, as this is the only therapeutic option available. It is extremely unlikely that these patients will develop pulmonary vascular disease precluding surgical intervention.

References