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Case Report

The anomalous drainage of the inferior vena cava into the left atrium

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ABSTRACT

Background. Anomalies in systemic venous return most commonly involve a persistent left supe-rior vena cava draining into the left atrium. Anomalous drainage of the inferior vena cava (IVC) into the left atrium is a rare congenital vascular disorder. The diagnosis was confirmed as anoma-lous drainage of the right superior pulmonary vein and large atrial septal defect following echo-cardiography. Anomalous drainage of the inferior vena cave was confirmed with computed tomog-raphy (CT). We report a rare combination of drainage of the inferior vena cava associated with atrial septal defect (ASD) and partial anomalous pulmonary venous return.

Case. A 14-year-old girl was referred to our hospital for the evaluation of palpitations, hypoxia, exertional dyspnea, and cyanosis. Transthoracic echocardiography (TTE) revealed a large sinus venosus ASD and anomalous right superior pulmonary venous return. A cardiac CT demonstrated IVC drainage to the left atrium and an anomalous right superior pulmonary vein draining into the right atrium.

Conclusions. In older patients with cyanosis, further imaging methods together with TTE will be useful in detecting additional cardiac anomalies. Patients with inferior vena cava opening to the left atrium are different from caval type ASD's and should be surgically repaired using a patch. Corrective surgery involves repositioning of the interatrial septum via a patch.

Key words: inferior vena cava, left atrium, anomalous pulmonary venous drainage.

Anomalies in systemic venous return most commonly involve a persistent left superior vena cava draining into the left atrium (LA). Anomalous drainage of the inferior vena cava (IVC) into the LA is a rare congenital vascular disorder. It has been reported in isolation and in association with other cardiac defects.[1] It can occur with atrial septal defect (ASD), anomalous pulmonary venous drainage, and pulmonary arteriovenous fistula.[2] In this case report, we described clinical signs and symptoms which include hypoxia, exertional dyspnea and cyanosis. Diagnosis confirmed anomalous drainage of the right superior pulmonary vein and a large ASD following echocardiography. Anomalous drainage of the IVC was confirmed with computed tomography (CT). We report a rare combination of drainage of the IVC associated with ASD and partial anomalous pulmonary venous return.

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Case Report

A 14-year-old girl was referred to our hospital for the evaluation of palpitations, hypoxia, exertional dyspnea, and cyanosis. Her blood pressure was 100/60 mmHg, and her heart rate was 85 beats/min. Oxygen saturation was 94% at rest in room air. An electrocardiogram (ECG) showed normal sinus rhythm and right ventricular hypertrophy. Transthoracic echocardiography revealed a large sinus venosus ASD and anomalous right superior pulmonary venous return. A cardiac computed tomography demonstrated IVC drainage to the LA and an anomalous right superior pulmonary vein draining into the right atrium (Fig. 1). There was no evidence of other cardiac

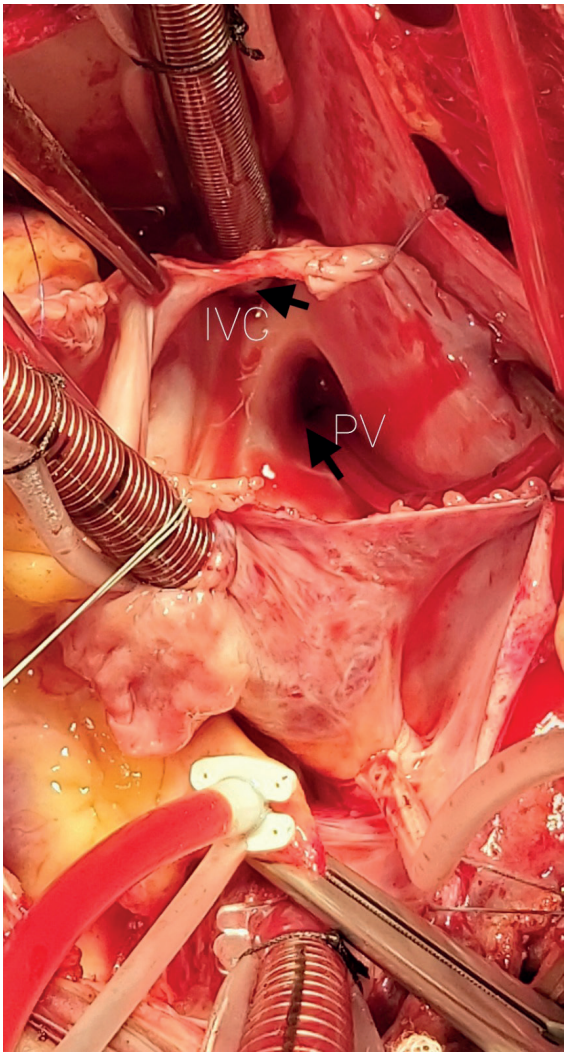

Fig. 1. Abnormal right superior pulmonary vein flowing into the left atrium and right atrium is shown.

anomalies. Informed parental written consent was taken for case publication.

The operation was performed through a median sternotomy incision. The ascending aorta and both vena cavae were cannulated directly. IVC cannulation was made as low and as close to the diaphragm, as possible. After initiated cardiopulmonary bypass under moderate hypothermia, the heart was arrested by antegrade del-nido cardioplegia. The right atrium was opened, which revealed a large ASD, 4x3 cm in size, and the inferior left-ward shift of IVC and anomalous drainage

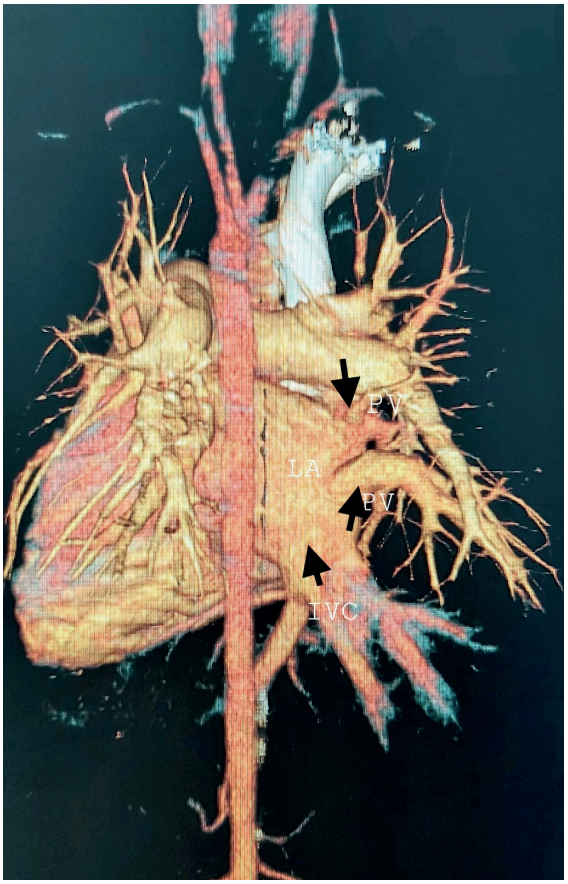

Fig. 2. Intraoperative pulmonary vein appears to open into the inferior vena cava.

of the right superior pulmonary vein (Fig. 2). The defect was closed with a patch of the fresh autologous pericardium. The IVC was redirected to the right atrium, and the right superior pulmonary vein was redirected to the LA. The postoperative course was uneventful, with the patient followed up fully saturated in room air. She was discharged on the fifth postoperative day.

Discussion

Anomalies in systemic venous return have been reported extensively, but drainage of the IVC directly into the LA is a rare condition, resulting in significant right-to-left shunts. However, anomalous drainage of the IVC into the LA is a rare congenital vascular disorder and is less commonly associated with an ASD

which Gardner first described in 1955.[1] An ASD occurring with the condition is uncommon in the reported cases. Anomalous pulmonary venous drainage and pulmonary arteriovenous fistula may also be associated.[2] This entity is different from a low or IVC secundum ASD shunting of blood from the IVC to the LA. Most patients with IVC drainage to the LA are diagnosed either congenitally or after incorrect ASD repair. If the surgeon is not careful, this can be mistaken for the inferior ASD rim, and they may iatrogenically divert IVC blood to the LA upon ASD closure.[3,4]

Patients clinically present most commonly at a young age with symptoms of right-to-left shunt. The main clinical features are shortness of breath, cyanosis, and palpitations. However, these congenital findings have been reported in asymptomatic patients referred for the evaluation of incidentally noted hypoxemia or cyanosis.[5]

Only a few cases with anomalous drainage of the IVC into the LA have been reported in the literature. Diagnosis can be difficult, as can be understood from the fact that most of the cases reported in the literature are diagnosed in adulthood.[5] More advanced imaging methods are often required to make a diagnosis. Here we present a case of a 14-year-old girl with ASD combined with anomalous drainage of the left superior pulmonary vein and drainage of the IVC into the LA. In our case, the TTE failed to demonstrate the anomalous IVC drainage into the LA. However, the presence of cyanosis made us suspect an associated disorder then the diagnosis was established by CT contrast angiography. On imaging, the IVC is positioned normally in the lower chest but then curves towards and joins the LA. In older patients with cyanosis, further imaging methods together with TTE will be useful in detecting additional cardiac anomalies. Patients with IVC opening to the LA are different from caval type ASD's and should be surgically repaired using a patch. Corrective surgery involves repositioning of the interatrial septum via a patch.

Ethical approval

Informed parental written consent was taken for case publication.

Author contribution

The authors confirm contribution to the paper as follows: study conception and design: AA, ANE; data collection: ANE; manuscript preparation: AA, ANE. All authors reviewed the results and ap-proved the final version of the manuscript.

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Conflict of interest

The authors declare that there is no conflict of interest.

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