Delayed presentation of partial anomalous systemic venous connection

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ABSTRACT

Anomalies of systemic venous connections are extremely rare. We describe the case of an asymptomatic 29-year-old woman who was found to have systemic desaturation in the peripartum period and referred to us for suspected cyanotic heart disease. She was diagnosed to have hemianomalous systemic venous connection of the inferior vena cava (IVC) into the left atrium (LA). Transesophageal echocardiogram with contrast diagnosed anomalous connection of the IVC to the LA, further confirmed by computed tomography and conventional angiography. The patient underwent successful surgical correction with an uneventful postoperative course.

Keywords: Anomalous drainage, cyanosis, inferior vena cava, systemic vein abnormality

INTRODUCTION

It is unusual for cyanosis to come to medical attention for the first time in adulthood. Pulmonary arteriovenous malformations, methemoglobinemia, and anomalous systemic venous drainage to the left heart are some of the conditions which might have such a late presentation in an otherwise asymptomatic adult. We report a case of anomalous connection of the inferior vena cava (IVC) to the left atrium (LA) in association with partial anomalous pulmonary venous drainage and an ostium secundum atrial septal defect (ASD).

CLINICAL SUMMARY

An asymptomatic 29-year-old female was suspected to have congenital heart disease due to cyanosis detected during pregnancy. Her pregnancy and parturition were nevertheless uneventful, and she sought cardiac evaluation several months postpartum. On examination, she was cyanosed and clubbed, with Spo2 88% in room air which dropped to around 80% following exertion. The cardiac apex was diffuse. On auscultation, she had a wide and fixed second heart sound with a loud pulmonary component and a grade II ejection systolic murmur at the left upper sternal border. Chest X-ray showed cardiomegaly with a right ventricular apical contour and dilated hilar branch pulmonary arteries. Transesophageal echocardiogram (TEE) showed a dilated right heart, a large ASD (with a predominant left to right shunt) and normal opening of the coronary sinus ostium into the right atrium (RA) [Supplementary Videos 1 and 2]. The bicaval view was difficult to obtain and the IVC could be traced, connecting to the LA. The Eustachian valve was not prominent and saline contrast study done from the lower limb showed opacification of the LA, followed by RA opacification through the ASD [Figure 1a and Supplementary Video 3]. Cardiac catheterization showed a drop in saturation from the pulmonary vein to

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the LA [Table 1]. IVC angiogram showed its connection to a posteriorly located LA, followed by opacification of the RA [Figure 1b]. Computed tomography (CT) confirmed the same, while the right superior vena cava connected normally to the RA [Figure 2a-d]. A large fossa ovalis defect with partial anomalous drainage of the right upper pulmonary vein (RUPV) to RA was also noted. In view of high pulmonary artery pressures and resistance [Table 1], she was planned for fenestrated closure of the ASD, rerouting of the RUPV to the LA and baffling of the IVC to the RA. The preoperative findings were confirmed intraoperatively [Figure 3]. Post repair, pulmonary artery pressures were one-third of the systemic pressures. Postoperative recovery was uneventful and she was discharged on the 6th postoperative day. She is asymptomatic at her last follow-up visit, 3 years later, with no features of pulmonary hypertension and unobstructed IVC flows to RA and RUPV flows to LA [Figure 4a and b].

Table 1: Cardiac catheterization data

Site	Saturation (%)	Pressure
SVC	64	
IVC	71	
Right atrium	83	a6v5m5
Right ventricle	-	76/6
Pulmonary artery	83	74/17/36
Pulmonary vein	99	a6 v5 m5
Left atrium	89	a6 v5 m5
Left ventricle	-	106/6
Aorta	91	113/70/84
Qp=6.4 L/min	-	-
Qs=3.8 L/min	-	-
PVRI=6.25 WU-m^2	-	-

SVC: Superior vena cava, IVC: Inferior vena cava, PVRI: Pulmonary vascular resistance index

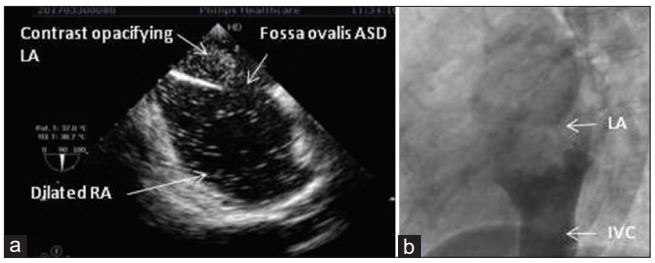

Figure 1: (a) Transesophageal echocardiogram with agitated saline contrast injected into a vein in the lower limb, densely opacifying the left atrium. The dilated right atrium is opacified via the atrial septal defect. (b) Left lateral angiogram showing a posteriorly placed inferior vena cava draining to the posteriorly located left atrium, adjacent to the spine

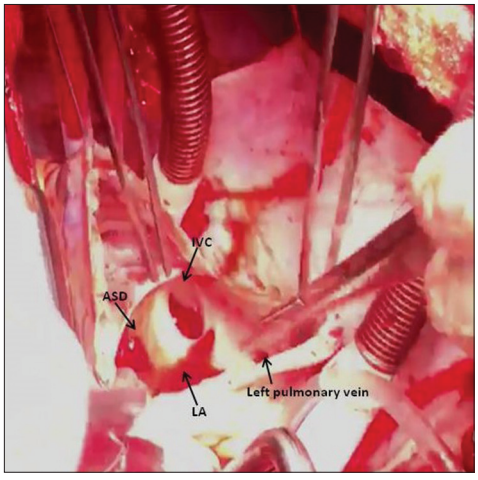

Figure 3: Intra-operative photograph demonstrating the anatomic peculiarities of the case

DISCUSSION

Most case reports of anomalous drainage of the IVC to the LA have described an abnormally large right valve of

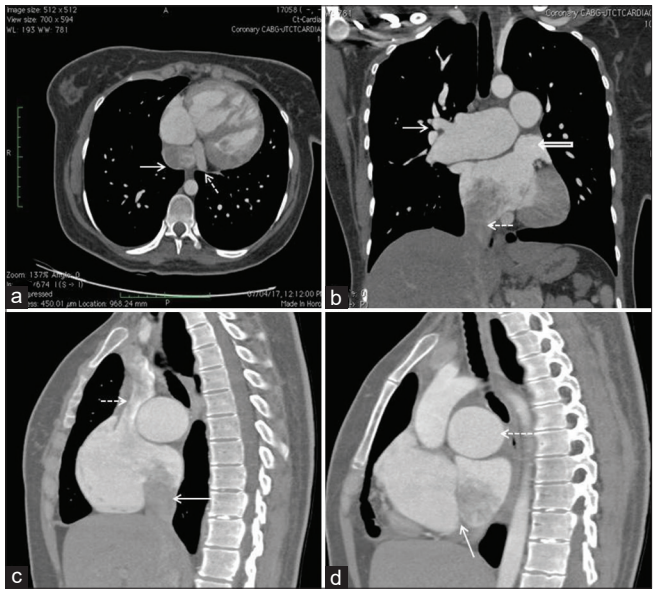

Figure 2: (a) Axial computed tomography section - Contrast washout in the left atrium due to anomalous inferior vena cava connection (arrow). Normal position of the coronary sinus (dotted arrow). (b) Contrast computed tomography (coronal plane) - Right middle pulmonary vein (arrow) connecting normally to the left atrium, anomalous inferior vena cava entry to left atrium (dotted arrow) and the left atrium appendage (bold arrow). (c) Contrast computed tomography (sagittal plane) - Normal drainage of right superior vena cava to right atrium (dotted arrow) and posteriorly located inferior vena cava to left atrium (arrow). (d) Contrast computed tomography (sagittal plane) - Plane of the inter atrial septum (arrow) and enlarged right pulmonary artery (dotted arrow)

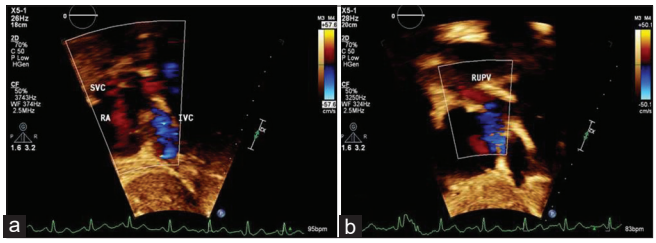

Figure 4: (a and b) Postoperative subcostal bicaval view showing the inferior vena cava baffled to right atrium and right upper pulmonary vein to left atrium with an intact atrial septum

the sinus venosus - the Eustachian valve - coexisting with an ASD or patent foramen ovale.[1,2] The persistence of a large Eustachian valve in various forms is a recognized entity and can lead to right to left streaming across an interatrial communication. During early embryologic development, the right and left venous valves separate the systemic sinus venosus from the primary part of the RA. The left venous valve subsequently fuses with the developing atrial septum. As the systemic sinus venosus becomes incorporated into the RA, the right venous valve diminishes. Rudiments are found in the crista terminalis, the valve of the IVC (Eustachian valve) and the valve of the coronary sinus (Thebesian valve).[3]

The heart as seen in ancestor fish consisted of a sinus venosus, a common atrium, a ventricle and an outflow tract. Complete separation of the systemic and pulmonary circulations, as seen in mammals and birds, necessitated the formation of a right ventricle and the development of an atrial septum, separating a right atrial half (receiving the systemic venous return from the body), from a left half (receiving oxygenated blood from the lungs via the pulmonary veins). Our case had anomalous connection of the IVC and not just anomalous drainage. This was well demonstrated on TEE and confirmed intraoperatively. The sinus venous, as early as the 5th week of development, receives the vitelline, cardinal and umbilical veins. The terminal portion of the IVC is formed from the right vitelline vein. Recent lineage analysis has demonstrated a separate origin of the myocardium of the systemic and pulmonary venous tributaries.[4] While the sinus venosus develops at the junction of the splanchnic and somatic mesoderm, the pulmonary veins develop from a splanchnic venous plexus which extends from the heart to the liver.[5] The pulmonary veins after losing connection with the splanchnic plexus draining to the liver eventually connect to the LA. The embryonic pulmonary vein is a midline structure connected to a common atrium, flanked by a so-called right and left pulmonary ridge. It eventually drains to the LA because the primary atrial septum starts to grow from the right pulmonary ridge, by which the midline pulmonary entrance becomes a "left atrial entrance." This is achieved by atrial septation, which begins from the right pulmonary ridge. The fact that part of the pulmonary veins drained incorrectly to the RA, along with improper drainage of the IVC to the LA, suggested that the molecular guidance of the venous returns was impaired and not the heart, *per se*.[6] This is corroborated by the fact that the coronary sinus drained normally into the RA, suggesting that the left sinus horn and its wall had developed normally and maintained their normal relation in the atrioventricular groove. The large ASD might have resulted from molecular misguiding in this area, leading to an abnormal position or impaired growth of the secondary atrial septum.

A systematic segmental echocardiographic approach, aided by saline contrast, confirmed the diagnosis. A second imaging modality using angiography, CT and/or magnetic resonance imaging, is helpful prior to surgery. While a lower pulmonary vascular resistance is a prerequisite for ASD closure, cyanotic pretricuspid shunts like total anomalous pulmonary venous connection often have a favorable outcome after surgical re-routing despite the presence of slightly higher pulmonary vascular resistance. Our case had a pretricuspid component (ASD with partial anomalous pulmonary venous drainage of the RUPV) and a posttricuspid component (IVC to the LA). Cyanosis and the accompanying polycythemia contribute to elevated pulmonary vascular resistance due to a combination of hypoxemia, altered endothelial function, and microthrombi formation. Relief of systemic desaturation often leads to reversal of these factors and a corresponding fall in the pulmonary pressure and resistance after successful surgical repair. Rerouting of the IVC back to the RA also prevents future cerebrovascular accidents, which these patients are increasingly prone to. Singh *et al.* reported a similar case, but their patient had persistent cyanosis after repair of a fossa ovalis defect.[7] Surgical repair in our case consisted of baffling the IVC and anomalous right pulmonary vein and simultaneous patch closure of the ASD [Supplementary Videos 4 and 5]. Since postoperative pulmonary artery pressures were normal, a fenestration in the ASD patch was not considered necessary by the surgeon. The pulmonary pressures have stayed normal during a recent follow-up visit [Supplementary Video 6].

CONCLUSIONS

We present a rare congenital malformation involving anomalous connection of the IVC into the LA. A certain degree of hypoxemia is well-tolerated permitting normal survival to adulthood, often with minimal symptoms. Cyanosis may be completely correctable by surgery, in these instances.

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Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand

that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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