Total Anomalous Systemic Venous Return to Right-Sided Atrium with Left Atrium Morphology: A Case Report

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Abstract

A 14-month-old boy with cyanosis, clubbing, left hemiparesis and convulsions was diagnosed by contrast echocardiography and angiocardiography to have anomalous connection of the venae cavae into the physiologic left atrium as the cause of his manifestations, in addition to situs inversus totalis and multiple small left to right shunts. It is important to consider this rare anomaly in the differential diagnosis of cyanosis and systemic emboli ***(Iranian Heart Journal 2004; 5(4): 61-64).***

Key words: venae cavae ■ anomalous connection ■ situs inversus totalis ■ congenital heart disease

The anomalous connection of the venae cavae into the anatomic left atrium (or the physiologic left atrium in situs abnormalities) is a very rare anomaly. The number of reported cases in the English literature is under 20.[2] Cyanosis and paradoxical embolic events are often the representing features. We have diagnosed this anomaly in a patient using peripheral contrast echocardiography and angiocardiography.

Case Report

A 14-month-old patient was referred to our clinic with mild cyanosis since birth, and early clubbing and left hemiparesis for one month. He had a history of convulsions before the beginning of his hemiparesis, brought under control by phenobarbital. His cardiovascular examination revealed mild cyanosis, early clubbing, palpable peripheral pulses in all four limbs, normal heart sounds and a grade III/VI systolic murmur at his right sternal border.

He had a hemoglobin level of 10.5 g/dL and a hematocrit of 35%. O2 saturation was 75%. There were no Howell-Jolley bodies in the peripheral blood smear.

The EKG showed inverted p-waves in leads III and AVf, and left superior QRS wave axis. His X-ray films suggested visceral situs inversus. There was dextrocardia with increased cardiothoracic ratio and peripheral vascular markings, and pulmonary artery prominency (Fig. 1).

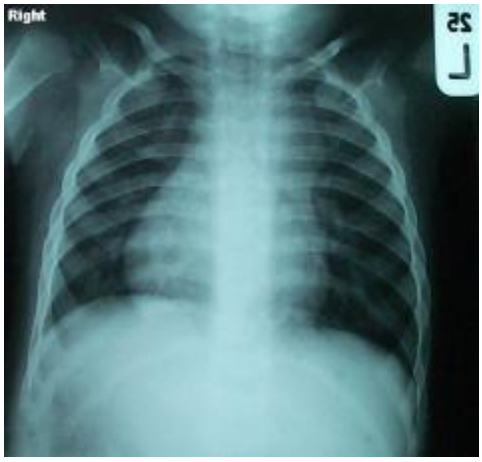

Fig. 1. Chest X-ray.

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Abdominal ultrasonography showed a unique right-sided spleen.

Standard echocardiography examination was performed using a VingMed CFM 750 echocardiograph with a 3.25 MHz transducer. The infrahepatic inferior vena cava (IVC) was not seen. Pulmonary veins drained into the right-sided atrium. The left-sided atrium was small.

There was a small atrial septal defect (ASD). No venous drainage to the left-sided atrium was visualized. The right-sided atrium drained into the right-sided ventricle with left ventricular morphology and thence into the aorta. The small left-sided atrium drained into a small left-sided ventricle with right ventricular morphology and thence into the pulmonary artery (PA). A small perimembranous ventricular septal defect (VSD) and a small patent ductus arteriosus (PDA) were seen.

The aorta was on the left side and anterior to the PA and arched to the right. The flow across semilunar and atrioventricular valves was normal except for trivial mitral and tricuspid regurgitation and a 35mmHg pressure gradient across the pulmonary valve [pulmonary valvar stenosis (PS) or flow gradient]. The right-sided ventricular ejection fraction was about 70 percent.

As we could neither explain the cyanosis of our patient nor show drainage of his SVC (superior vena cava), we performed a peripheral contrast echocardiography. We used agitated saline as a contrast agent, injecting it into right and left upper extremity veins via intravenous lines. The contrast material first appeared in the right-sided atrium, which drained into the aorta via the right-sided ventricle at a point near the mitral valve (Fig. 2).

To confirm the diagnosis, we performed angiocardiography. The results of pressure and O2 saturation measurements are seen in Table I. The catheter was inserted into the right femoral vein and thence, via a right-sided vein (azygos), it was passed into the SVC. From there, it passed easily into the atrium and then into the ventricle. Injection at that site yielded the result that the ventricle was right-sided with left ventricular morphology and that it directly oppacified the aorta, although PA was also opacified via PDA and VSD (Fig. 3).

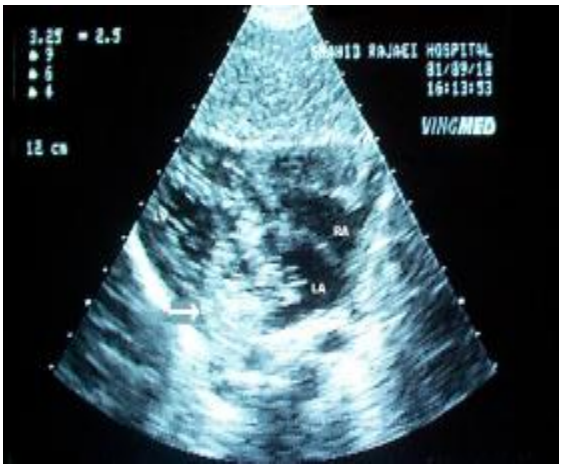

Fig. 2. Contrast echocardiography, subcostal view. LA: physiologic left atrium (right-sided); RA: physiologic right atrium (left-sided); LV: morphological left ventricle (right-sided); Arrow = the place where agitated saline first appeared. The left-sided chambers are hypoplastic.

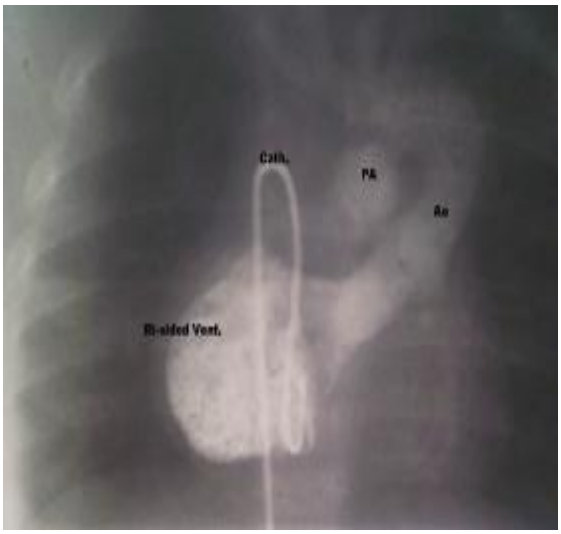

Fig. 3. Angiocardiography, injection into right-sided ventricle. Cath: catheter, PA: pulmonary artery, Ao: Aorta, Rt-sided Vent.: right-sided ventricle.

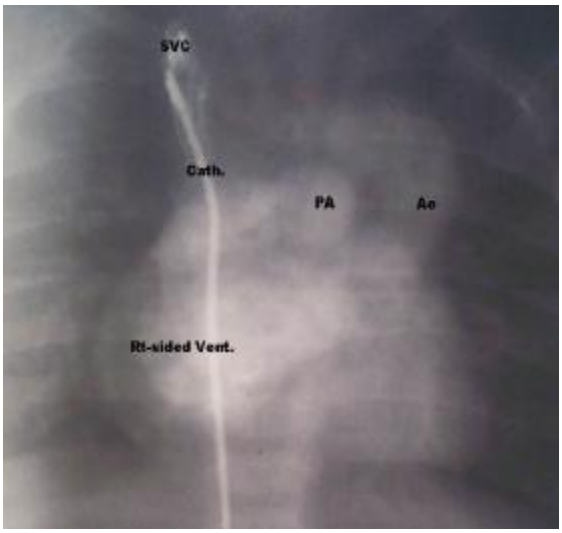

Fig. 4. Angiocardiography, injection into superior vena cava. SVC: superior vena cava, Cath.: catheter, PA: pulmonary artery, Ao: aorta, Rt-sided Vent.: right-sided ventricle.

Injection into the SVC confirmed its abnormal drainage to the systemic side of the heart (Fig. 4). The final diagnosis for our patient was: situs inversus totalis, dextrocardia, L-loop, L-malposition of the aorta, small ASD, small perimembranous VSD, small PDA, interrupted IVC with azygos continuation, anomalous connection of venae cavae to the right-sided physiologically left atrium, mild PS or pulmonary valve flow gradient, and right aortic arch.

Table I: Catheterization data

Site	Pressure (mmHg)	O2 Saturation (percent)
SVC		47
Right-sided atrium	12/5 (mean=8)	74
Right-sided ventricle	100/5	75
Femoral artery	100/60 (mean=80)	75

The patient was referred to a surgeon for total correction of his anomaly, if possible. The surgeon confirmed drainage of venae cavae and pulmonary veins to the right-sided atrium with left atrial morphology and the presence of small left-sided chambers. As total correction was not possible due to the small sizes of those chambers and in order to decrease the patient's cyanosis, an atrial septectomy was performed in the hope that it would increase the left-sided chamber flow for total correction in the future. After the operation, O2 saturation rose to 89% and there was a 12mm post-surgical ASD at echocardiography.

Discussion

The anomalous connection of venae cavae to the systemic side of the heart is a very rare anomaly. It often presents with cyanosis and paradoxical embolic events, causing diagnostic challenges due to its rarity. The total number of reported cases in the English literature is under 20. Interestingly, this case is the second one from our center.[5] Although this hospital is the major referral center for complex congenital heart disease patients from Iran and some neighboring countries, it seems that the occurrence of two cases of this very rare anomaly could not be explained by chance alone.

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