Case Report

Anesthetic implications of total anomalous systemic venous connection to left atrium with left isomerism

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ABSTRACT

Total anomalous systemic venous connection (TASVC) to the left atrium (LA) is a rare congenital anomaly. An 11-year-old girl presented with complaints of palpitations and cyanosis. TASVC with left isomerism and noncompaction of LV was diagnosed after contrast echocardiogram and computed tomography angiogram. The knowledge of anatomy and pathophysiology is essential for the successful management of these cases. Anesthetic concerns in this case were polycythemia, paradoxical embolism and rhythm abnormalities. The patient was successfully operated by rerouting the systemic venous connection to the right atrium.

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INTRODUCTION

Total anomalous systemic venous connection (TASVC) is a rare congenital anomaly. It may be seen in isolation or with other congenital anomalies. There may be situs solitus or cardiac malpositions. It may also be part of heterotaxy syndrome.[1]

Anesthetic management of TASVC and heterotaxy syndrome requires detailed assessment of cardiac and extracardiac anomalies and thorough knowledge of its pathophysiological effects. We report anesthetic management of a case of TASVC to the left atrium (LA), with left isomerism.

CASE REPORT

An 11-year-old female patient weighing 25 kg presented with a history of fatigue for 2 years, cyanosis for 2 months and occasional palpitations. There was no history of squatting or cyanotic spells. There were no symptoms suggestive of congestive heart failure. On examination, pulse rate was 60/min, BP was 100/60 mmHg, jugular venous pressure was not elevated and room air oxygen saturation was 80%. Cardiovascular examination revealed normal first heart sound and single second heart sound and a grade 2/6 ejection systolic murmur. Her hemoglobin was 18.6 g/dL, with hematocrit of 51.42%. She had serum bilirubin of 1.3 mg/dL and serum creatinine of 0.8 mg/dL. ECG showed p wave appearing after QRS complex and no evidence of left or right ventricular hypertrophy. On chest X-ray, there was no cardiomegaly, pulmonary vascular markings were normal and both domes of diaphragm were at the same level. Preoperative echocardiogram showed that the patient had situs solitus, osteum secondum atrial septal defect (ASD), left to right shunt and interrupted inferior vena cava (IVC). Superior vena cava (SVC) was draining to left-sided atrium, There was an enlarged LA, noncompaction of left ventricle (LV) and normal biventricular function. Contrast echocardiogram delineated the entry of agitated saline into left-sided atrium from SVC [Figure 1]. Cardiac catheter course was from SVC to left-sided atrium and then to right

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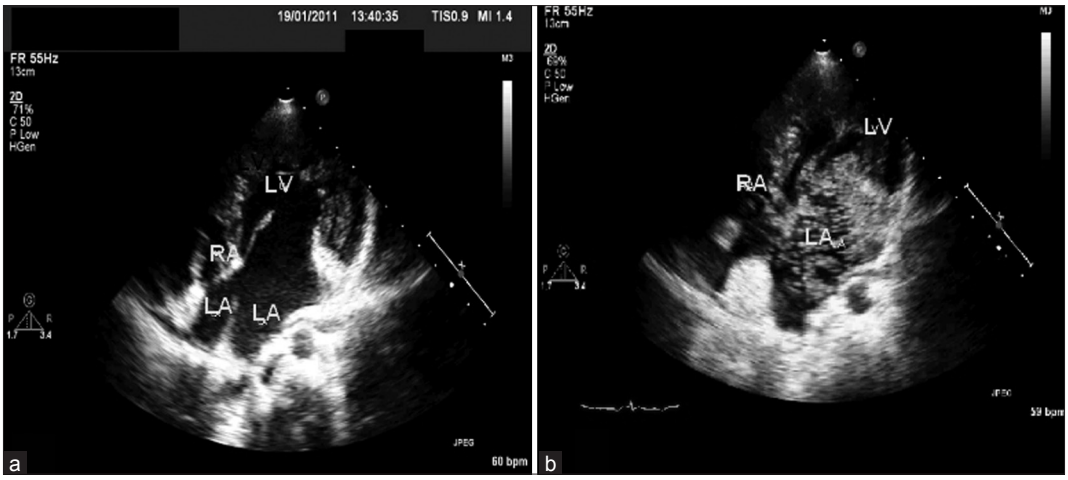

Figure 1: (a) Transthoracic four-chamber view showing dilated left atrium (LA), left ventricle (LV), noncompacted LV, a membrane dividing LA into two parts and atrial septal defect (ASD) with a small right atrium (RA). (b) Contrast ECHO showing contrast entering LV and RA from LA

atrium (RA), right ventricle (RV) and the pulmonary artery. Pulmonary venous saturation was 99%, there was desaturation at LA (80%), and no further change in the saturation was found at RA, RV or PA.

Computed tomography (CT) angiography was performed to confirm venous anomalies, and diagnosis of left isomerism was made with polysplenia and bilateral left-sided bronchi, midline liver. There was interruption of the IVC with azygous continuation, with the hepatic vein joining the LA separately [Figure 2]. After the investigations, the patient was diagnosed as a case of TASVC and left isomerism.

Preanesthetic concerns were about polycythemia, altered coagulation profile and rhythm abnormalities. Goals of anesthesia were prevention of air embolism and to avoid drugs known to precipitate arrhythmias and to avoid drugs known to cause myocardial depression as non-compacted LV may be predisposed to myocardial dysfunction and intravenous bolus injections reach coronary circulation faster in TASVC.

On the day of surgery, the patient was premedicated with tab diazepam 5 mg 1 h before surgery. Preoperatively, hydration was maintained by IV fluids. In the OR, the radial arterial blood pressure was 100/60 mmHg and pulse rate was 60/min. General anesthesia was induced with midazolam 2 mg, ketamine 50 mg and glycopyrrolate 0.2 mg. Endotracheal intubation was done after muscle relaxation with vecuronium. Right internal jugular vein was cannulated with a 7 French double-lumen central venous catheter. Anesthesia was maintained with isoflurane 0.5–1% in oxygen–air 50%, and boluses of fentanyl 25 mcgs and vecuronium 1 mg. During prebypass period, care was taken to avoid air embolism. Once the pericardium was opened, the preoperative findings were confirmed [Figure 3]. Anticoagulation was achieved with heparin 100 mg and ACT was 782 s. Ascending aorta, SVC and suprahepatic IVC draining the hepatic veins were cannulated and cardiopulmonary bypass (CPB) initiated. Cold blood cardioplegia was given through aortic root after applying the cross-clamp. Left-sided atrium was opened and there was a membrane dividing the same into two parts, which was initially confused to be the atrial septal defect. Left side of the membrane was receiving pulmonary veins and, on right side of the membrane, SVC, hepatic vein and coronary sinus openings were noticed. The RA was rudimentary, which was opened through right-sided atrial appendage. Tricuspid valve was inspected and was of adequate size. Atrial septal defect was enlarged, membrane in the LA was excised and the SVC, hepatic vein and coronary sinus were rerouted to the right atrial cavity with baffle repartitioning using pericardial patch. The RA was enhanced with pericardial patch and closed. Total bypass time was 116 min and aortic cross-clamp time was 92 min. Cardioplegia was repeated twice. The minimum temperature on CPB was 28°C. CPB was conducted with a flow of 2.4 L/min/m.^{2} After rewarming to 36°C. Dopamine 5 mcg/kg/min was started electively and the patient was weaned from CPB without any difficulty. Protamine was given after achieving surgical hemostasis. Postoperatively, hemoglobin was maintained at 10 g/dL. Arterial saturation was 100% after surgery. The patient was weaned from the ventilator and extubated after 4 h in the recovery room.

Postoperative echocardiogram showed an intact interatrial patch, no residual ASD, SVC and hepatic veins with suprahepatic IVC draining to RA. Biventricular function was normal.

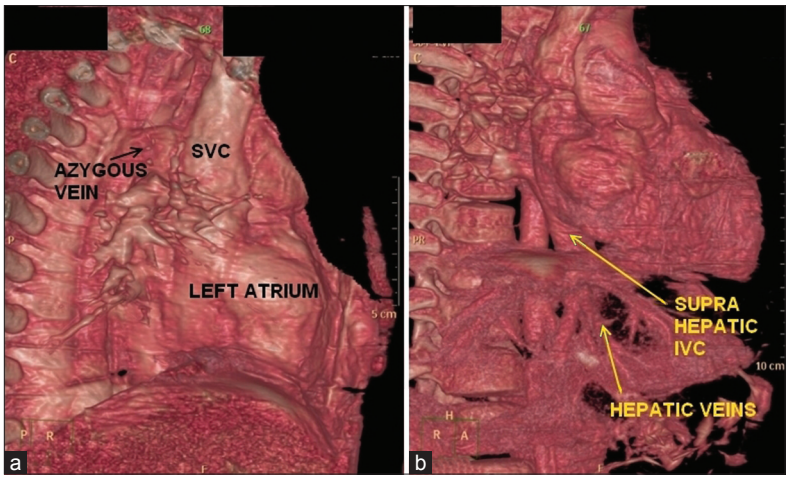

Figure 2: Computed tomography angiogram showing (a) azygous vein draining into the enlarged superior vena cava (SVC) and (b) hepatic veins draining into the suprahepatic inferior vena cava (IVC)

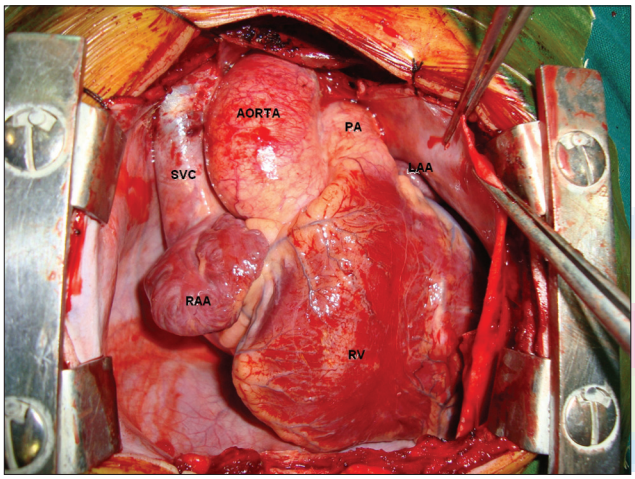

Figure 3: Peroperative view after sternotomy showing morphological right atrial appendage (RAA) appearing as a finger-like projection (left atrial isomerism) and engorged superior vena cava (SVC) draining into left atrium (LA)

DISCUSSION

Anomalous systemic venous drainage consists of anomalies of SVC, IVC, and TASVC. Persistent left SVC is the most common systemic venous anomaly, whereas there are less than 20 cases of TASVC reported in the world literature.

When all the systemic venous return is to the LA, presence of an ASD is essential for survival. Physiologic consequences of TASVC vary from asymptomatic to severe cyanosis, depending on the size of the interatrial communication; larger the defect less the desaturation. Pulmonary blood flow (PBF) is determined by the volume of mixed pulmonary and systemic venous blood crossing the ASD to reach the RA, RV and pulmonary circulation. In these patients, the RV may be normal or hypoplastic because right-sided venous return may be reduced.[2]

Maape *et al*.[2] reported a case of TASVC with bilateral SVC and hypoplastic RA and RV. Diagnosis of TASVC was done when the child was 4 months old. The child presented with cyanosis without episodes of hypoxic spells. The patient was operated at the age of 15 years when she developed dyspnea on exertion. Complete surgical correction was not possible as RV was hypoplastic and hence bilateral bidirectional Glenn and rerouting of IVC and coronary sinus to RA were performed.

TASVC patients are prone to paradoxical embolism. In a case report by Mogadam *et al*.,[3] a cyanotic child with left hemiparesis and convulsions was diagnosed to have TASVC by contrast echocardiogram and angiocardiography. Schick EC Jr *et al*.[4] reported a case of anomalous SVC that was diagnosed incidentally during pulmonary artery catheter insertion after acute myocardial infarction. The patient was acyanotic and did not undergo surgery for anomalous SVC. He died of brain abscess 4 years later. They recommended surgical correction of such conditions even if patients are asymptomatic. Ze-Wei *et al*.[5] classified TASVC into two types. In type I, IVC is not interrupted and in type II, there is IVC interruption. Miller *et al*.[6] and Gueron *et al*.[7] reported children between the ages of 3 and 15 years with diagnosis of TASVC who were successfully operated by rerouting the systemic venous drainage to RA.

Marcelletti *et al*.[8] described two cases of TASVC, one child with right isomerism and double-outlet RV and pulmonary stenosis underwent palliative shunt but died postoperatively whereas another child with left isomerism and partial atrioventricular septation

who underwent anatomical repair survived. In our case, TASVC was associated with left isomerism. Left isomerism or bilateral left-sidedness is associated with bilateral left atrial appendages, bilateral morphologic left bronchi and bilateral morphologic bilobed left lungs. Superior vena cavae are bilateral and attach to morphologic left atria. In left isomerism, SA node may be absent or hypoplastic. Fetal bradycardia and complete heart block may help to diagnose fetal left isomerism. Atrial pacemaker is ectopic in the atrial wall or near the coronary sinus ostium (coronary sinus rhythm) due to persistence of embryonic left-sided pacemaker. There may be sinus bradycardia, junctional rhythm or heart blocks, ranging from first degree to complete heart block. Abnormal automaticity of atrioventricular node may be the cause of junctional ectopic tachycardia seen after correction of congenital cardiac anomaly.[9] Our patient presented with complaints of palpitations. ECG showed left axis deviation of the p wave, with p waves appearing after QRS complex. Perioperatively, the child was closely watched for arrhythmias. On the fifth postoperative day, although the patient complained of palpitations, there was no change in the rhythm and palpitation did not require treatment. Extracardiac gastrointestinal malformations in the left isomerism are polysplenia, transverse liver, intestinal malrotation, biliary atresia, duodenal/esophageal atresia and congenital short pancreas. Clinical course of left isomerism is determined by cardiovascular anomalies and extracardiac malformations and not because of polysplenia.[1]

CONCLUSIONS

TASVC is a cause of cyanosis and clubbing in patients with no other signs of heart disease. TASVC draining only through the SVC to LA with interrupted IVC and noncompaction of LV is a rare association. When TASVC is part of heterotaxy syndrome, detailed evaluation of all the associated malformations is required. Attention should be paid to preoperative hydration and relief of anxiety, avoiding air embolism. Intravascular volume should be maintained and pulmonary vascular resistance should not be allowed to rise. Noncompaction of LV, which was an associated feature in our patient, can lead to LV dysfunction. It requires avoiding cardiodepressant drugs and thorough deairing before removal of the cross-clamp. Perioperatively, patients with left isomerism should be closely monitored for arrhythmias.

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