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Left sided Superior Vena cava: (A case report and review of literature)

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Abstract

A male child aged about one and half year old was referred to the department of Radio diagnosis for CT vena cavography after he was examined on 2-dimensional echocardiography which revealed the diagnosis of Fallot's tetralogy. On CT scan besides the confirmation of presence of Tetralogy, there was an abnormal finding of left sided superior vena cava, an uncommon association.

Keywords: Persistent left superior vena cava; 64 slice CT; developmental malformation

1. Case History

One and a half year old male child presented to the outpatient department with the chief complaints of breathlessness, cough, fever and features of central cyanosis and pan digital clubbing since three months of age. He had history of repeated respiratory tract infections and hospitalized many times for the same complaints without diagnosis of this entity. On examination the heart rate was 143 beats / minute and respiratory rate 28 / minute. No bony chest deformity. Per abdominal examination showed mild hepatomegaly. On Echocardiography he was diagnosed to have Tetralogy of Fallot (TOF), with classical features of sub-infundibular pulmonary stenosis, ventricular septal defect, Right ventricular hypertrophy and over-riding of aorta. Then the patient was referred to us for CT confirmation and vascular study of cardiac haemodynamics.

Under proper sedation, the child was scanned on GE Light speed VCT 64 slice CT scan covering the area from thoracic inlet to diaphragm. After scanogram, the non-ionic contrast media (about 35ml) was injected through bilateral antecubital vein, images were acquired in axial planes in 0.625mm collimation and the diagnosis was ascertained. Two days later a separate scan was again obtained through left sided antecubital vein. Axial and coronal scan were obtained using MIP (Maximum intensity Projection) and volume rendering soft ware technique. The additional findings to Tetralogy to Fallot's was seen with presence of left sided superior vena cava (SVC) besides right sided superior vena cava, the normal observation. The left SVC was opening through dilated left coronary sinus into the right atrium (Fig. 1, 2 & 3).

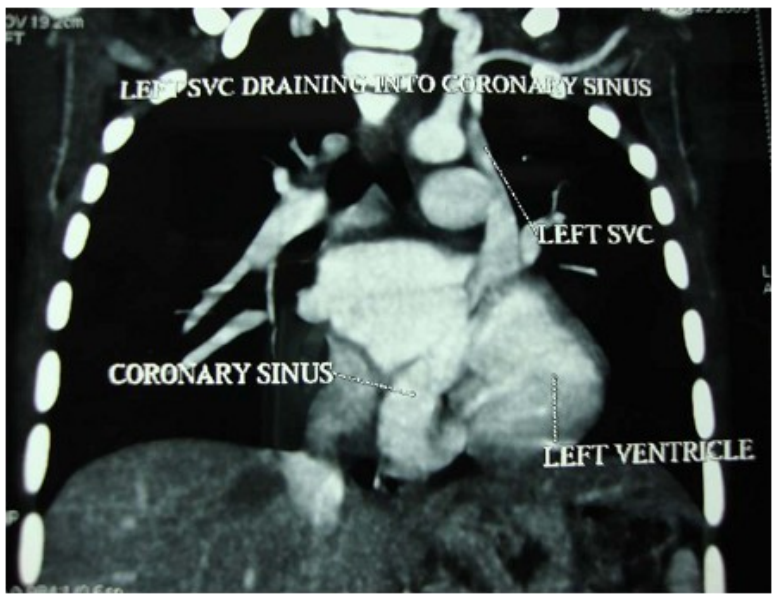

Fig 1.Coronal MIP image showing Left SVC coursing into posterior AV ring to drain into coronary sinus of right atrium.

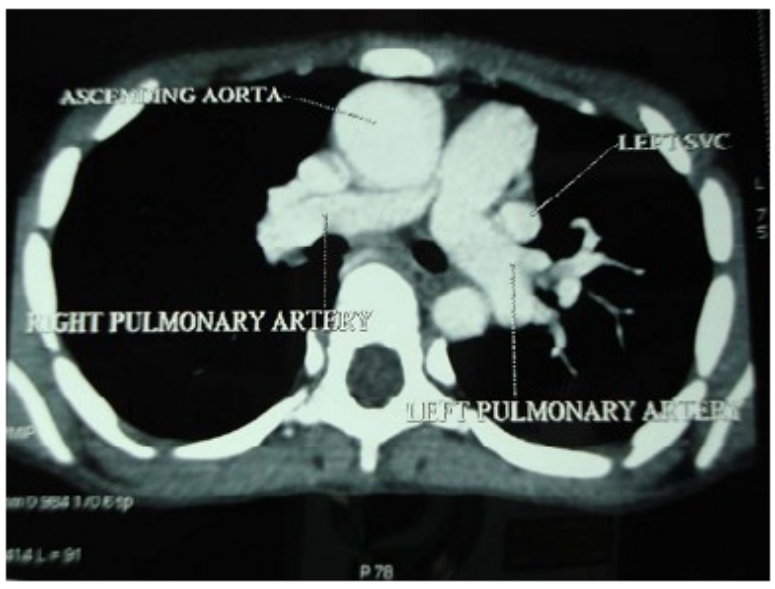

Fig 2. Axial CT image showing double SVC on either side of Arch of Aorta with the Left SVC coursing left of main pulmonary trunk towards posterior AV groove.

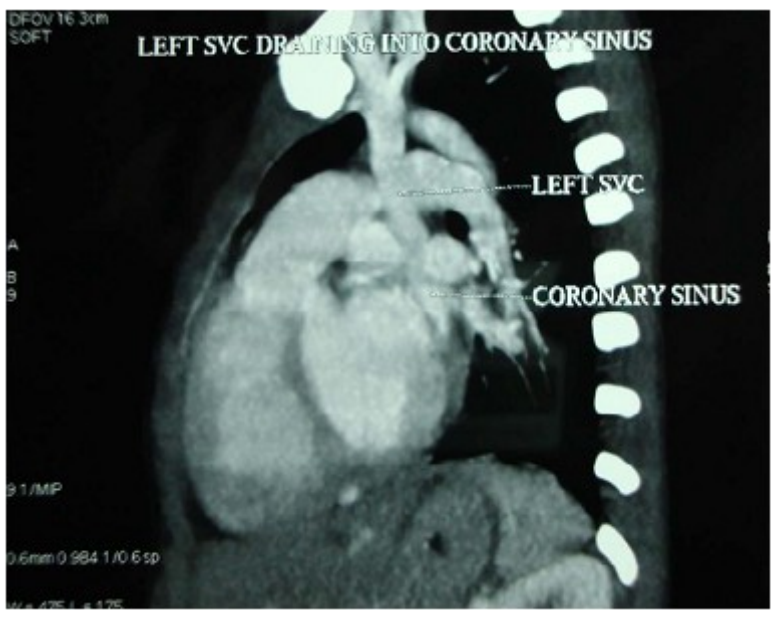

Fig 3. Sagittal MIP image showing the course of Left SVC and its drainage into dilated coronary sinus.

No other cardiac as well as pulmonary abnormalities could be detected. And thus finally a diagnosis of TOF with left sided superior vena cava was entertained. The TOF was surgically corrected. Post operative period in the hospital was uneventful. The patient was called for follow up every two months.

2. Discussion

Persistent left superior vena cava (SVC), a congenital venous anomaly, occurs in 0.5% of general population, 0.3% of healthy population and 4.3% of the patient with congenital heart diseases.[1] In 82% of the patients with Left SVC a right sided SVC is also present. Persistence of Left SVC with, absent right superior vena cava occurs in only 0.09 – 0.13% of patients with congenital heart disease.[2]

This anomaly is frequently associated with situs inversus. Persistent left SVC with other anomalies and structures in the left atrioventricular groove may be overlooked if they are not carefully and appropriately evaluated.

Persistence of left cardinal vein (which normally obliterates during embryogenesis) results in persistent left SVC.[3] Persistent left SVC itself does not produce any physiological derangements but is associated with other congenital heart disease like septal defects, Tetralogy of Fallot's, situs inversus etc.[4] Left SVC drains either into right atrium via coronary sinus or into left atrium. 92% of Left SVC drains into right atrium via left portion of sinus venosus or coronary sinus and are asymptomatic. They are considered to be anomaly of coronary sinus. 8% of left SVC drains into left atrium and causes right to left shunt and may present with unexplained cyanosis and clubbing. It is a problem to keep blood out of the field during cardiopulmonary bypass and thus the left SVC may need to be ligated or separately cannulated to avoid its venous return causing distension of right heart. Also it is a contraindication to retrograde cardioplegia.[5] Other problems may include arrhythmia cardiac arrest and coronary sinus thrombosis. It can

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cause difficult left sided central insertion of pulmonary artery catheter or pacing wire attributable to orientation.[3] The clinical signs include jugular venous distension on the left side, abnormal left jugular venous waveform due to direct transmission of left atrial contraction, atrial pressure and non-arterial blood gas analysis.

Conventional chest X-ray usually display widening of aortic shadow, para median bulge along left heart border and crescentic vascular shadow projecting from left upper border of aortic arch to the middle third of the clavicle.

Other diagnostic tools are venography without fluoroscopy[7], transthoracic echocardiography and multislice CT scan or dual source CT scan. Safest diagnostic option is trans- thoracic echocardiography with an agitated saline microbubble contrast media which has the advantages of being non invasive, without radiation effect and causing less haemodynamic instability in a sick person compared with contrast imaging.[5] It also demonstrates drainage of contrast into the coronary sinus.[8] It is likely to be associated with heterotrophy (discordance or malposition of thoraco-abdominal organs and vessels, complex congenital heart anomalies and extra cardiac defects or defect involving midline structures. It could be associated with interruption of inferior vena cava and splenic anomalies. It can present like left sided paracardiac mass due to dilated coronary sinus. The diagnosis is usually confirmed on multislice row detector CT scan.

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