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PERSISTENT LEFT SUPERIOR VENA CAVA DRAINING INTO THE LEFT ATRIUM – A CASE REPORT

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Contribution

RT conceived idea, did literature review and final drafting. SK & AK reviewed case report. All authors contributed significantly to the submitted manuscript.

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ABSTRACT

Twenty two years young female presented with shortness of breath, palpitations and paroxysmal nocturnal dyspnoea (PND) in outpatient department. Her Past history of rheumatic fever was positive. She had irregular pulse, heaving apex beat and raised JVP. Pansystolic murmur present in mitral area and axilla. ASOT was 300 but TLC was normal. Echocardiogram showed severe mitral regurgitation. She was planned for mitral valve replacement. Intraoperative transesophageal echocardiography (TEE) showed persistent left superior vena cava (PLSVC) that was opening into the left atrium. TEE findings of PLSVC opening in LA were confirmed as excessive blood was flowing in LA after opening the left atrium. A vent was placed in LA due to excessive blood flow. We also put a suction catheter and were then able to proceed further and the mitral valve then replaced. Patient was discharged on warfarin and digoxin in healthy and stable state.

Key Words: Persistent left superior vena cava, Superior vena cava, Mitral regurgitation, Left atrium

INTRODUCTION

Persistent left superior vena cava (PLSVC) is the most common venous congenital anomaly of thoracic cavity. [1] It has been reported in 0.4% cases in normal population and 5% of those with congenital heart diseases. It is formed due to persistence of left anterior cardinal vein during fetal development.

Mostly it opens in right atrium through coronary sinus and is of no practical implications. It is also asymptomatic when it drains in left atrium, but when combined with hemodynamic instability or raised RA pressure, it can cause right-to-left shunting and hypoxia.[2] Cyanosis does not occur in symptomatic patients because the shunt is usually not large enough since it only drains the left upper limb and left side of the head and neck.

CASE REPORT

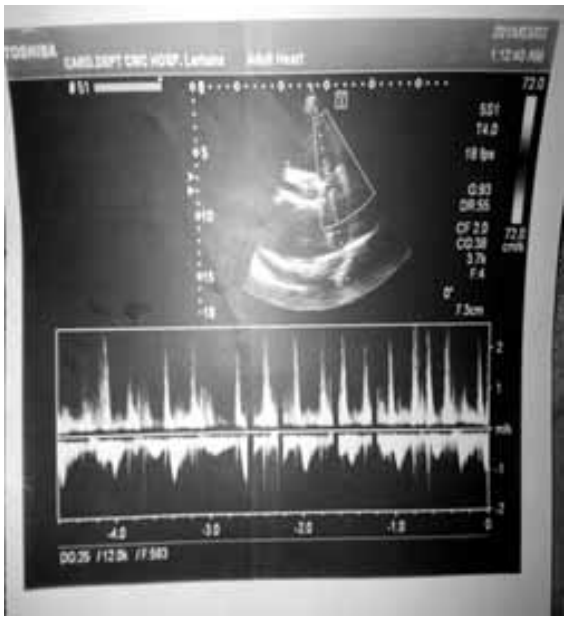
A 22 years old female presented in May 2015 in OPD of department of Cardiac Surgery, Civil Hospital Karachi with shortness of breath (NYHA class III),

palpitations and paroxysmal nocturnal dyspnea. Past history was positive for rheumatic fever.

On examination, she was anemic. Pulse was irregular. Apex beat was palpable in left 5th intercostal space lateral to midclavicular line and was heaving in nature. Pansystolic murmur was heard in mitral area and was radiating to axilla. Jugular venous pressure was raised. There was no carotid bruit. Rest of the systemic examination was unremarkable. Her preoperative workup was positive for anemia with a Hb of 9.6 g/dl. ASOT was positive and was 300 but total leucocyte count was within normal limits. Rest of the labs were also within normal limits.

Transthoracic echocardiogram showed severe mitral regurgitation with thickened mitral valve. Left atrium was dilated with size of 60mm. Ejection fraction was 64% with moderate to severe pulmonary arterial hypertension (PAP 65mmHg) (Figure 1).

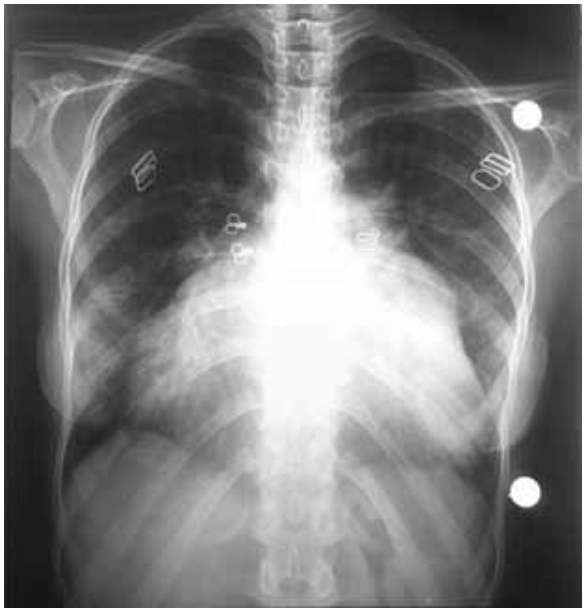
Figure 1: Transthoracic Echocardiogram of the Patient Showing Mitral Regurgitation



Preoperative chest x-ray (PA view) showed cardiomegaly, with no other positive finding on x-ray (Figure 2). She was diagnosed as case of severe mitral regurgitation and planned for mechanical mitral valve replacement with preservation of posterior leaflet. After taking informed consent for surgery as well as case report, she was prepared for surgery. In operation theatre, on air ABGs were done which showed pH of 7.47 at FiO_2 of 100%. PO_2 was 85.5mmHg and PCO_2 was 34.6mmHg. HCO_3level was 26.4mmol/L and base excess was of 1.8mmol/L.

Under general anesthesia, invasive and non-invasive monitoring was done. Transesophageal Echocardiogram (TEE) was performed which showed persistent left superior

Figure 2: Chest X-Ray PA View of the Patient Showing Cardiomegaly



vena cava (PLSVC) that was opening into the left atrium. Median sternotomy was done and standard cardio-pulmonary bypass established with aortic and bicaval cannulation. We found right as well as left SVC and both were of approximately equal sizes. Persistent left superior vena cava (SVC) was opening into the left atrium. After cross clamping the aorta, cold blood antegrade cardioplegia was given through aortic root needle. Left atriotomy was then done.

After left atriotomy, a vent was placed in the left atrium but due to excessive dark colored blood coming into the LA through the opening of PLSVC, we put a suction catheter into the opening of PLSVC that was opening into the LA. Mitral valve was then assessed, and anterior mitral leaflet excised while posterior leaflet preserved and the valve was replaced with double disc mechanical valve. Left atriotomy closed with proline, continuous stiches and de-airing done and CPB weaned off smoothly. Hemostasis secured, mediastinal drain placed, pacing wire inserted, wound closed in layers and patient shifted to ICU with minimal ionotropic (Adrenaline) support.

In ICU on ventilator-spontaneous (SMV) mode ABGs showed a normal pH of 7.39 at 70% FiO_2. PO_2 was 100mmHg and PCO_2 was 36.7mmHg. She was maintaining 100% saturation. HCO_3 level was 22.9mmol/L.

Patient was weaned off from ventilator smoothly. After extubation ABGs were normal and showed a pH of 7.44 at 21% FiO_2. PO_2 was 90.1mmHg but she was maintaining 100% saturation. PCO_2 was 34mmHg. Clinically she was stable with no sign of cyanosis.

On 2nd postoperative day, she was shifted to the ward and was discharged on 5th postoperative day on warfarin and digoxin. She was advice to take precaution for swan-Ganz catheter placement, CVP placement and pacemaker implantation if required in the future. Her first OPD visit was after one week. On that and the successive visits she was stable haemodynamically.

DISCUSSION

Persistent left superior vena cava (PLSVC) is the most common venous congenital anomaly of thoracic cavity. [1] It has been reported in 0.4% cases in normal population and 5% of those with congenital heart diseases. It is formed due to persistence of left anterior cardinal vein during fetal development. Mostly it opens in right atrium through coronary sinus (92%) and is of no practical implications. It is also asymptomatic when it drains in left atrium, but when combined with hemodynamic instability or raised RA pressure, it can cause right-to-left shunting and hypoxia.[2]

Our patient had both left SVC and right SVC of approximately equal sizes. Persistent left SVC was draining into the left atrium but there was no hypoxia, cyanosis and the patient was also hemodynamically stable.

The most common subtype of PLSVC results in the presence of both left and right SVCs. A right sided SVC may be normally present (82-90% of cases). Left brachiocephalic vein is also seen as a persistent bridging vein in 25-35% of cases.[3] Webb *et al* reported that a PLSVC is associated with absence of the innominate vein in 65% cases. More rarely, the caudal right superior cardinal vein regresses leading to an absent right SVC with PLSVC.

In our patient, there was right SVC and also a left SVC. Both SVCs were of adequate sizes. But, there was no bridging innominate vein. PLSVC may be detected in fetal life. C Berq et al state that fetal anatomy should be scanned closely as PLSVC may be found associated with other anomalies. However, isolated PLSVC is a benign anomaly which usually does not affect the outcome.[4,5]

Our patient had no associated congenital heart defect but she had irregular pulse and isolated severe mitral regurgitation. Past history was positive for rheumatic fever. Radiological investigations to confirm PLSVC include Echocardiogram (dilated coronary sinus and Bubble Study), Chest Xray (if a catheter or line is in an unexpected left paramediastinal location) or CT Chest. Nuclear Medicine, Transesophageal Echocardiography and Radionuclide angiocardiography are also used.[6-8]

We performed chest x-ray (PA view) and transthoracic echocardiography. Both modalities were inconclusive for the presence of left SVC. Intraoperative transesophageal echocardiogram (TEE) was performed which showed persistent left superior vena cava opening in left atrium.

PLSVC may become significant in cases of venous procedures such as line placement or pacemaker implantation.[3] Significance of PLSVC increases in permanent pace maker, implantable cardioverter defibrillator and when left subclavian vein is used to access the right side of the heart or the pulmonary vasculature. Swan-Ganz catheter is widely used but inserting it can be tricky. Presence of PLSVC becomes challenging during cardiac surgeries when retrograde cardioplegia is given.[7]

In our case we used only antegrade blood cardioplegia for myocardial protection. PLSVC usually follows a favorable prognosis except where a large right-to-left shunt is present. Our patient had favorable prognosis. Left SVC was draining into the left atrium without producing hypoxia or cynosis. Patient was hemodynamically stable in her six months follow up visits.

CONCLUSION

Persistent left superior vena cava (PLSVC) is a rare congenital anomaly which may produce difficulty during surgery but does not usually impact the outcome. Presence of PLSVC produces a difficult task when right side of the heart has to be accessed. In such a case Swan-Ganz catheter placement is usually recommended.

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