

Persistent left superior vena cava in patient with right atrial myxoma undergoing open heart surgery. A case report and review of literature

ABSTRACT

Persistent left superior vena cava (PLSVC) is a rare and asymptomatic congenital cardiovascular anomaly. Being asymptomatic, PLSVC was usually discovered while performing interventions (such as insertion of central lines, Swan-Ganz catheters, or placing pacemakers) through the left internal jugular vein or left subclavian veins. Commonly, PLSVC is detected not only as an isolated congenital anomaly, but also it can be associated with many other cardiac anomalies. Also, presence of a dilated coronary sinus on echocardiography should raise the suspicion of PLSVC. The diagnosis should be confirmed by contrast venography or computed tomography angiography. The present case is a female patient, 29 year old, who was undergoing elective excision of a right atrial mass, with closure of patent foramen ovale, and she had end-stage renal failure on regular hemodialysis three times weekly through a permicath inserted in the right subclavian vein.

Key words: Cardiac surgery; left superior vena cava; right atrial myxoma

Introduction

The incidence of persistent left superior vena cava (PLSVC) is generally rare (0.1–0.5% of the general population) and up to 10% of those with established congenital heart disease.^[1] However, it is actually the most common congenital anomaly of thoracic venous system.^[2] Embryologically, PLSVC results when the left anterior cardinal vein fails to obliterate.^[3]

Commonly, PLSVC can be detected as isolated congenital anomaly, but it can also be associated with other cardiac anomalies including tetralogy of Fallot (TOF), patent foramen

ovale (PFO), atrial septal defect (ASD), ventricular septal defect (VSD), bicuspid aortic valve, coarctation of aorta, coronary sinus ostial atresia, and cor triatriatum.^[4]

Echocardiography cannot detect the presence of PLSVC, but it can detect presence of a dilated coronary sinus, and this should raise the suspicion of PLSVC, and the diagnosis should be confirmed by magnetic resonance venography, multislice computed tomography (CT) but these are not routine prerequisites to cardiac surgery.^[5]

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How to cite this article: Elhamamsy M, Aldemerdash A, Zahran F, Bakir BM, Alanazi NA, Awadallah YA, *et al.* Persistent left superior vena cava in patient with right atrial myxoma undergoing open heart surgery. A case report and review of literature. Saudi J Anaesth 2020;14:217-20.

Access this article online	
Website: www.saudija.org	Quick Response Code 
DOI: 10.4103/sja.SJA_511_19	

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Submitted: 11-Aug-2019, **Accepted:** 24-Sep-2019, **Published:** 05-Mar-2020

Case History

A 29-year-old female was brought by her family due to nausea, vomiting, and restlessness for 3 days.

Past medical history of

(1) DM type I, since childhood on insulin, (2) ESRD (presumed to be 2ry to DM nephropathy), on regular hemodialysis through permicath inserted in the right subclavian vein, (3) HTN 4- hypothyroidism on levothyroxine.

Chest X-ray showed: No infiltration or signs of overload.

Patient was admitted to ICU.

2 days later: She developed weakness in her left arm, associated with difficulty in speaking with fluctuating aphasia (thalamic aphasia).

MRI brain: showed right thalamic acute/sub-acute lacunar infarction.

Neurology Consultation: recommended TEE examination.

TEE showed: A large clot in RA at tip of dialysis catheter, measuring 3.4×2.5 cm. There is evidence of PFO with evidence of left to right shunt.

Cardiac MRI showed:

Moderate to large right atrial mass ($32 \times 28 \times 25$ mm) originating close to IVC. Collectively, (DDx will be myxoma and less likely angiosarcoma).

CT Cardiac Findings: Right atrial mass with no extra cardiac extension.

Final decision was to proceed with cardiac surgery.

She was taken to the operating room for excision of the right atrial mass, and closure of PFO.

After anesthesia induction, and because of the presence of the permicath in RSCV, it was mandatory to insert the central venous access in left internal jugular vein (LIJV) to avoid any complications with the right atrial mass or the inserted permicath. So, a French 7 triple lumen central venous catheter, and Swan-Ganz sheath were inserted in LIJV using the anatomical landmarks, and it was very smooth without any complications. The central venous pressure (CVP) was monitored normally through the central venous catheter, and it was used also for administration of

drugs and sampling for activated coagulation time (ACT) without any events.

Postoperative Plan: The patient is transferred to SICU in stable condition.

Chest X-ray showed: The tip of the catheter is seen behind the left atrium [Figure 1].

Sample for blood gases was taken from the triple lumen catheter and the surprise was that it is typical as arterial and not venous sample, but, in the same time it displayed central venous wave and pressure.

Fluoroscopy showed the tip of the catheter behind the left atrium.

CT Chest with Contrast showed: Double SVC with persistent left SVC draining in left superior pulmonary vein. LIJV catheters tip is in PLSVC which drain to the left superior pulmonary vein.

This clarifies the arterial blood gases in samples from this catheter, and also CVP wave and pressure [Figure 2].

Surgical pathology report: Right atrial myxoma.

Discussion

Embryologically, PLSVC results from failure obliteration of the left anterior cardinal vein.^[3]

The thoracic embryonic venous system is composed of two large veins (the superior or anterior cardinal veins) which return blood from cranial aspect of embryo, and (the inferior

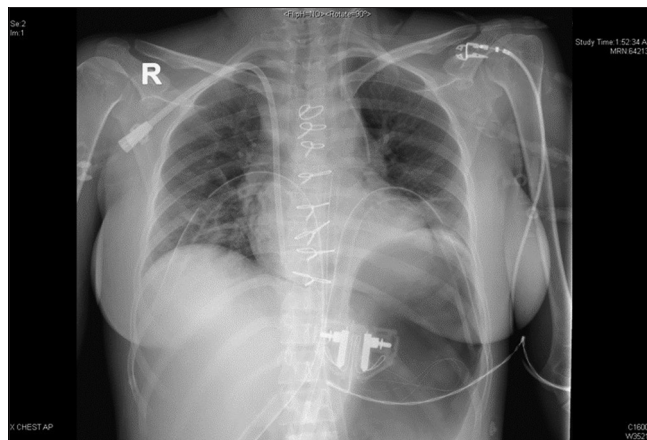


Figure 1: Postoperative chest X-ray showing the permicath inserted in the right subclavian vein, and the triple lumen central line inserted in the left internal jugular vein with the tip of catheter laying behind the left atrium



Figure 2: Postoperative chest CT angiography showing the central line inserted in persistent left SVC draining into the left superior pulmonary vein

or posterior cardinal vein, which returns blood from the caudal aspect.^[6]

Both pairs of veins join to form right and left common cardinal veins before entering the embryological heart. The left common cardinal vein persists to form coronary sinus and oblique vein of left atrium. The cephalic portion of superior cardinal veins form the internal jugular veins. The caudal portion of right superior vein forms the normal right-sided superior vena cava, while the portion of the left superior cardinal vein caudal to the innominate vein normally regresses to become “ligament of Marshall”.^[7]

Variations have also been reported in the insertion of PLSVC. In 80–90% of individuals, the PLSVC drains into the right atrium via the coronary sinus and is of no hemodynamic consequence [Figure 3].

In the remaining cases, it may drain in left atrium resulting in a right to left sided shunt.^[7]

In present case, PLSVC was draining in the left superior pulmonary vein [Figure 2], which is very rare anomaly, resulting in a left to right sided shunt, and sampling of the central line was typical arterial blood gases.

Detection of left SVC is usually made as an incidental finding during cardiovascular imaging or surgery.

In one case of PLSVC, placement of Swan-Ganz catheter via the left subclavian approach demonstrates an unusual course of the catheter on chest X-ray.^[5]

In another case, a 78-year-old male was undergoing elective placement of a permanent pacemaker. After achieving access through the LSCV, the wire kept on going on the left side of the chest instead of crossing the midline to the right side.

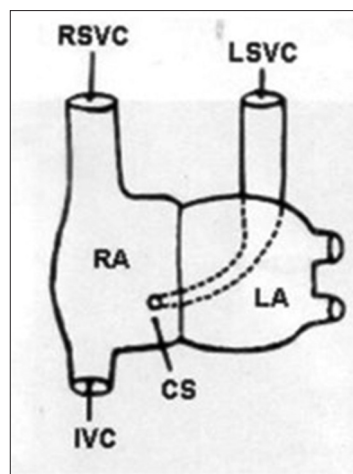


Figure 3: Diagram of left superior vena cava (LSVC) draining into the right atrium (RA) via coronary sinus (CS). The inferior vena cava (IVC) and right superior vena cava (RSVC) join the right atrium normally. LA = Left atrium. (Adapted from Cormier *et al.*,^[7])

The wire was removed and contrast venography was done, PLSVC with dilated coronary sinus emptying into the right atrium was confirmed.^[8]

For pacemaker placement, the greatest challenge is crossing the tricuspid valve to place the ventricular lead,^[9] which may rarely be complicated by dissection or perforation of the thin walled coronary sinus.

On the other hand, **during cardiac surgery**, the presence of PLSVC is a relative contraindication to the administration of retrograde cardioplegia. It may be possible to clamp the PLSVC to prevent the cardioplegia solution from perfusing retrograde up the PLSVC and its branches with inadequate myocardial protection.^[10] During heart transplantation in a patient with PLSVC, the coronary sinus must be dissected carefully to permit reanastomosis of PLSVC to right atrium.^[11]

Conclusion

During routine cardiac surgery and cardiological interventions, whenever a catheter or guide wire inserted via LJV or LSCV takes an unusual left-sided downward course, physicians should consider presence of PLSVC.

In patients with PLSVC, there are no contraindication to use LJV or LSCV for insertion of catheters or permanent pacemakers; however, there may be some technical difficulties and additional associated risks, and this should be discussed with the patient preoperatively; however, for safety, an alternative access sites should be considered whenever possible.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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