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Paradoxical emboli from left superior vena cava causing recurrent brain abscess

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ABSTRACT Persistent left superior vena cava is a rare but well-recognised condition. We describe a case of persistent left superior vena cava draining directly into the left atrium, with a fixed anatomical right-to-left shunt and paradoxical embolic events causing recurrent brain abscess. Surgical ligation was curative.

Keywords: brain abscess, left superior vena cava, stroke
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INTRODUCTION

Persistent left superior vena cava (SVC) occurs in 0.3%–0.5% of the population. It is associated with an increased incidence of congenital heart disease.[1] When it presents alone, it is asymptomatic. SVC is usually an incidental finding during cardiovascular imaging or surgery. A persistent left SVC can cause problems during surgical procedures such as a pacemaker implantation, central venous catheterisation and cardiopulmonary bypass surgery.[1] About 10% of persistent left superior vena cava drains directly into the left atrium, thus creating a right-to-left shunt and paradoxical emboli.[2]

CASE REPORT

A 53-year-old woman with a past medical history of epilepsy presented to the neurology department with an acute onset of right-sided weakness associated with worsening of her eczema over the preceding few days. She had a similar episode 15 years ago, and diagnosis of a brain abscess was made on computed tomography (CT) then. Physical examination revealed low-grade fever and peripheral cyanosis with 90% oxygen saturation on room-air pulse oximetry. There was no finger clubbing. Neurological examination indicated right-sided weakness with a power of 3/5. The patient also had pruritic excoriations over both her upper limbs. Blood investigation showed normal haemoglobin with leukocytosis, elevated erythrocyte sedimentation rate and C-reactive protein. Chest radiography was normal. Magnetic resonance imaging of the brain revealed a fairly well-defined right parietal lobe abscess with peripheral enhancement and extensive perifocal oedema. The abscess was drained and the patient completed a course of antibiotics.

In view of her recurrent brain abscess and peripheral cyanosis, we further examined the patient for signs of a right-to-left shunt. Chest CT revealed a persistent left SVC (Fig. 1), and transthoracic echocardiography revealed normal cardiac morphology. Agitated saline contrast injection via the right arm did not reveal an intra-cardiac shunt. Transoesophageal echocardiography (TEE) performed subsequently with agitated saline injected via the left arm revealed the anomalous SVC draining into the left atrium, above the left auricle. Venogram confirmed the left subclavian vein receiving the left internal jugular vein to form the left SVC (diameter = 1 cm) that drains directly into the left atrium (Fig. 2). There was no bridging left innominate vein to join the left SVC to right SVC.

Owing to the patient's recurrent brain abscesses, surgical closure was performed. Intrapericardial ligation of the left SVC was performed via a left anterior minithoracotomy. The left atrial appendage was identified with the left SVC that was draining into the roof of the left atrium (Fig. 3). Upon clamping the left SVC, the venous pressure rose by 6 mmHg. Triple ligation with transfixion of the left SVC was performed (Fig. 4). The patient had an uneventful recovery and has remained well with no recurrent brain abscess or seizures for the past three years.

DISCUSSION

The superior and inferior cardinal veins form the main drainage system of the embryo.[3] Both pairs of veins join to form the right and left common cardinal veins, before entering the embryological heart. The superior cardinal veins drain the cephalic part of the embryo and the inferior cardinal veins drain the rest of the embryo. At eight weeks' gestation, anastomosis between the right and left superior cardinal veins develops. During normal cardiac development, the right common cardinal vein forms the SVC. Anastomosis between the superior cardinal veins develops into the left innominate vein. The left common cardinal vein then disappears, leaving only the coronary sinus and the remnant ligament of Marshall. Persistence of the left cardinal vein results in formation of the left SVC. The bridging left innominate vein may or may not degenerate. In the vast majority of patients, the persistent left SVC drains into the right atrium via an enlarged coronary sinus. On rare occasions, the coronary sinus is absent,

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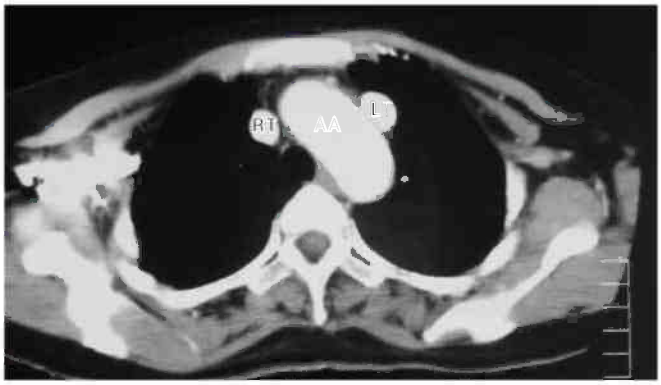

Fig. 1 Axial CT image shows a persistent left (LT) superior vena cava next to the aortic arch (AA), with a normal right (RT) superior vena cava.

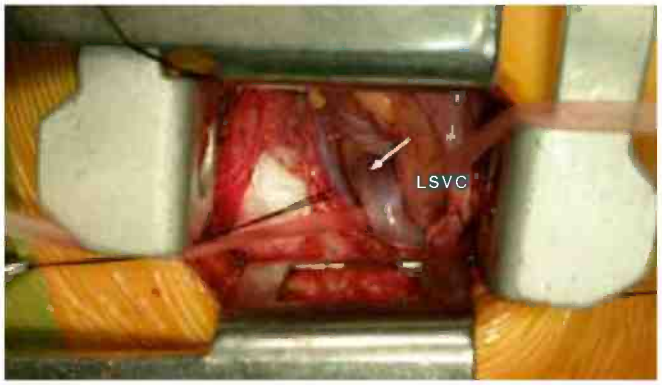

Fig. 3 Photograph shows a persistent left superior vena cava (LSVC) in a sling.

Fig. 2 Venogram shows a persistent left superior vena cava (LSVC) that drains directly into the left atrium (LA), resulting in a fixed-anatomical right-to-left shunt.

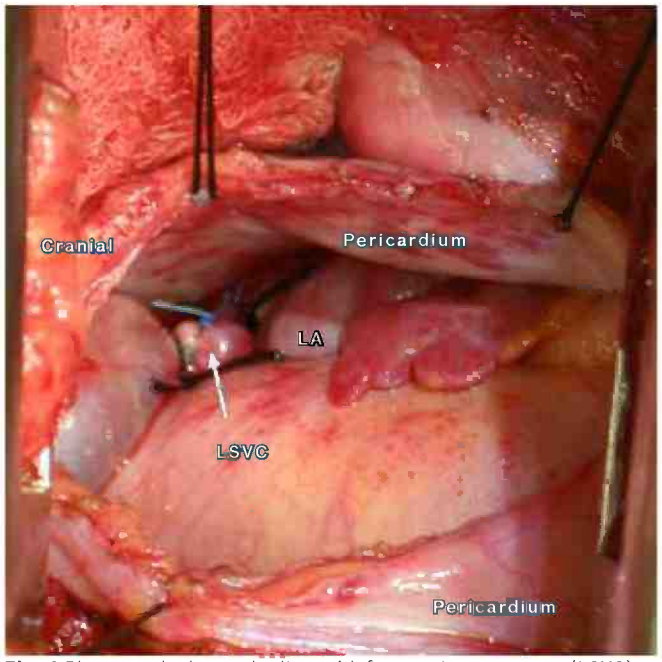

Fig. 4 Photograph shows the ligated left superior vena cava (LSVC) at the roof of the left atrium (LA).

and the persistent left SVC drains directly into the left atrium, resulting in a fixed anatomical right-to-left shunt.

This case highlights the importance of a thorough search for an anatomical right-to-left shunt in paradoxical embolic events, as this lesion may not be detected using routine investigation. In retrospect, a 'young' stroke at 38 years of age with peripheral cyanosis should have been a clue to our patient's diagnosis, even before the recurrence of her brain abscesses. Due to the strong clinical suspicion, TEE with contrast injection via her left arm had clinched the diagnosis. Her skin eczema was likely to be the infective source.

Several surgical procedures have previously been described for the management of persistent left SVC, including division and reimplantation of the left SVC to the right atrium, an intra-atrial baffle to divert flow from the left SVC to the right atrium, anastomosis of the left SVC to the left pulmonary artery and simple ligation of the left SVC.[4] Intracardiac rerouting techniques have been used traditionally to correct this right-to-left shunt. They are technically more difficult. Thus, complications are not uncommon and include prolong myocardial ischaemic time, early detachment and late deterioration of the baffle. Simple ligation of the left SVC may be the easier, safer and preferred procedure. Dural venous sinuses are communicating with each other. Left SVC can be safely ligated without causing cerebral oedema; the caveats being that the right SVC has to be of the same size or larger than the left SVC, and the venous pressure does not rise above 30 mmHg (normal venous pressure) after clamping.[4] During complete clamping of the left SVC, the mean venous pressure in the cephalic districts increases,[5] which may lead to venous hypertension and cerebral injury. The situation may be transient, as normal blood flow is usually restored in the presence of a normal right SVC. As less than 50% of patients are suitable for simple ligation, a left SVC pulmonary artery shunt may be an alternative in this situation. Endovascular approaches using coil and vascular plug to treat symptomatic persistent left SVC have been sporadically reported in the literature.[1,2]

This case highlights that persistent left SVC may not be 'benign' or important only to the cardiac surgeon dealing with congenital cardiac disease. A simple surgical correction without cardiopulmonary bypass is curative.

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