CASE REPORT

Catheterisation of a persistent left superior vena cava

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SUMMARY

We present a case of a 62-year-old woman with end-stage renal disease who displayed a persistent left superior vena cava detected following placement of haemodialysis catheter through the left internal jugular vein (IJV). The diagnosis was confirmed by left internal jugular vein cathetogram, computed tomography (CT) thorax and transthoracic echocardiography. The catheter was removed without immediate complications. She remained well during the outpatient follow-up.

BACKGROUND

Congenital anomalies of the superior vena cava are incidentally discovered during procedures which include central venous catheter (CVC) insertion, pacemaker electrode placement and cardiopulmonary bypass surgery.

Persistent left superior vena cava (PLSVC) is a rare anomaly found in about 0.3% of healthy subjects.¹ This condition results from failure of the left anterior cardinal vein to degenerate during embryological development. Serious complications such as shock, arrhythmias, angina and cardiac arrest have been described during catheterisation of adults with PLSVC.² Catheterisation of this vessel including haemodialysis catheter placement appears to be safe if its drainage into the right atrium is ensured.³ The main objective of this case report is to emphasise that any operator who is inserting a haemodialysis catheter in the left jugular or subclavian vein should be aware of the existence of PLSVC.

CASE PRESENTATION

A 62-year-old woman with end-stage renal disease (ESRD) and underlying ischaemic heart disease presented with a complaint of purulent discharge from her dialysis catheter site for 2 days. On clinical examination, she had a right neck collection from the infection.

She had been on regular haemodialysis via a right internal jugular CVC for over 2 months. She was empirically started on a course of intravenous ceftazidime 1 g daily and cloxacillin 1 g every 6 hours. A diagnosis of catheter-related bloodstream infection was made when a positive growth with *Corynebacterium striatum* was identified from both systemic and desired blood samples. The right internal jugular CVC was removed, and the antibiotic was changed to intravenous vancomycin 1 g daily. A left internal jugular CVC was inserted to continue the dialysis support. There was no immediate complication postinsertion of the CVC and

the arterial blood gases analysis confirmed venous blood. However, it was noted on the chest radiograph (figure 1) that the catheter was on the left side of the cardiac silhouette. There was no evidence of haemopneumothorax. Left internal jugular vein (IJV) cathetogram was performed and showed contrast flow from the left to the right atrium with no opacification of the aorta (video 1).

INVESTIGATIONS

CT scan of the neck and thorax was done to further evaluate the position of the left IJV CVC (figure 2). A transthoracic echocardiogram (TTE) demonstrated that the tip was not found in either of the ventricles or the atria, and most likely it was behind the left atrial appendage.

DIFFERENTIAL DIAGNOSES

When a CVC that is supposed to project into the superior vena cava abnormally courses to the left of the mediastinum, the following differential diagnoses should be considered; which could be the inadvertent catheterisation of the:

1. Left internal thoracic vein.
2. Left superior intercostal vein.
3. Left subclavian artery to the descending thoracic aorta.
4. Extravascular position into the mediastinum or pleural space.

MANAGEMENT

The PLSVC catheter was removed by the cardiothoracic team. There were no complications attributable to the catheter removal. Subsequently, she underwent uneventful placement of a permanent left femoral CVC for continuation of haemodialysis.

OUTCOME AND FOLLOW-UP

Once the anatomical anomaly was confirmed, the PLSVC catheter was discontinued and the patient had an uneventful hospital course. Her subsequent follow-up did not reveal any complications pertaining to the PLSVC.

DISCUSSION

PLSVC has an estimated incidence of 0.3%–0.5%.^{4,5} PLSVC occurs due to the failure of involution of the left cardinal vein during fetal life. It has been known that the presence of PLSVC is related to abnormal electrophysiological functioning of the heart. This is because of the anatomical and structural abnormalities of the conduction tissues and the sinus node (dilation of coronary sinus),^{6–8} resulting in

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Figure 1 Chest radiograph demonstrating the atypical course of the left central venous catheter. AP, anteroposterior.

bradyarrhythmias (atrioventricular conduction blocks) or tachyarrhythmias (supraventricular tachycardias, atrial fibrillation syndrome).

Most of the dialysis catheters are inserted through the right IJV, and right superior vena cava coexists with PLSVC in more than 80% of patients, hence rarely reported.^{9 10} PLSVC should be considered especially when the CVC via the left subclavian vein or IJV proved to be difficult. It is a known association with other congenital cardiac anomalies such as atrioventricular septal defect, ventricular septal defect and atrial septal defect.¹¹ In this case, there was no evidence of cardiac anomalies as shown by the TTE. In general, it is asymptomatic and clinically undetectable.^{12 13} Incidentally, PLSVC is typically detected during catheterising procedures, for example, in haemodialysis catheter placement. It can potentially lead to many serious complications, such as arrhythmias provocation, systemic embolisation, vascular thrombosis, shock, angina and cardiac arrest.²

Figure 2 CT scan revealed coronal mediastinal window with contrast showing the left-sided superior vena cava. The arrow showed the tip of the left internal jugular vein in the left superior vena cava.

Left superior vena cava may appear on the posteroanterior chest radiograph where it manifests itself as the widening of the aortic shadow, paramedian stripe, paramediastinal bulging or as a low density line along the upper left margin of the heart.¹⁴

It can be diagnosed non-invasively by echocardiography in conjunction with Doppler and/or contrast ultrasonography. CT scan and three-dimensional contrast-enhanced magnetic resonance venography are also important diagnostic modalities.⁶

A similar case was reported by Lim *et al*¹¹ when a catheterising procedure was performed under fluoroscopy. Initially they thought it was an arterial puncture. After confirming a venous puncture, they proceeded with the insertion of the catheter. A digital subtraction angiography revealed the PLSVC, and the patient was haemodialysed using the catheter for 5 months without any complications.⁷ Jang *et al* described another case when they had to insert a left-sided temporary dual lumen catheter on a patient with fistula failure, and the chest radiograph showed that the catheter tip was on the left paramediastinal side (similar to our case).⁸ The catheter was not removed immediately. After confirming the location of the catheter by CT scan, the diagnosis of PLSVC was made. Another case of PLSVC was reported by Stylianou *et al*,¹⁵ which was diagnosed incidentally after replacement of the haemodialysis catheter via the left IJV. Abnormal position of the catheter tip in the left paramediastinal area was revealed by the chest radiograph. CT scan and TTE were done to establish the diagnosis. She had been using the catheter for dialysis for a month before she was able to proceed with the newly fashioned graft.

With the rise in the number of patients with ESRD on haemodialysis in Malaysia, it is essential for clinicians who insert CVC to be aware of this congenital anomaly to prevent further complications. Moreover, it is vital to exclude other concomitant cardiac anomalies such as septal defects to prevent systemic embolism following thrombosis of the CVC.

Video 1 Left internal jugular vein cathetogram shows contrast flow from the left to the right atrium with no opacification of the aorta.

Learning points

- ▶ Patent left superior vena cava (persistent left superior vena cava (PLSVC)) has been diagnosed in 0.3%–0.5% of the general population.
- ▶ It results from the persistence of the left cardinal vein that usually involutes in later life.
- ▶ Serious complications such as shock, angina, cardiac tamponade and cardiac arrest have been described during catheterisation in adults with PLSVC.
- ▶ When difficulty arises on central venous catheterisation via the internal jugular or subclavian vein, variations of the superior vena cava should be entertained.
- ▶ It is paramount to ascertain the drainage pattern of PLSVC with venogram, echocardiography and CT prior to long-term catheterisation.

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