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Case Report

Surgery Section

Persistent Left Superior Vena Cava

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ABSTRACT

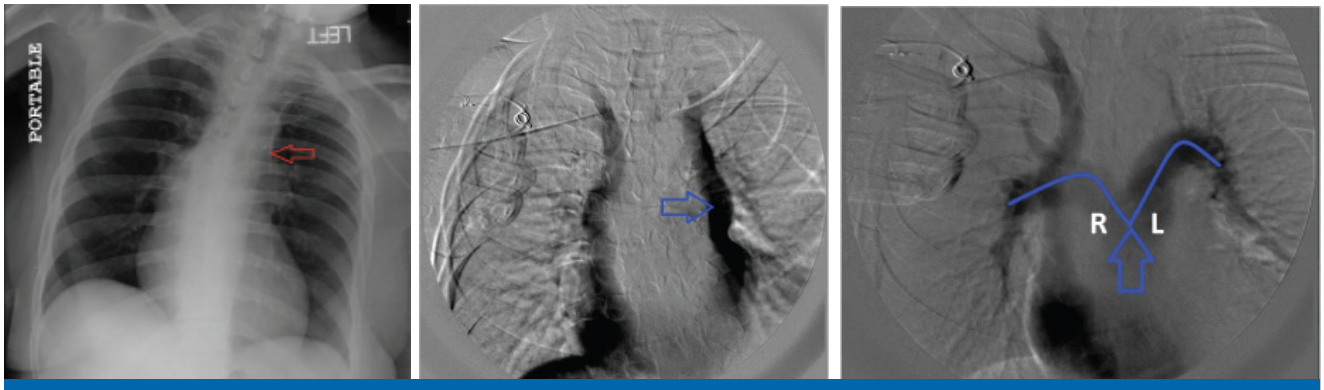
Persistent left superior vena cava is a rare anatomic anomaly, found in 0.3-0.5% of the general population, and upto 12% of patients with a congenital cardiac anomaly. Typically found incidentally, familiarity with such an anomaly can help clinicians to avoid complications during placement of central lines. This report describes a case of a patient with persistent left superior vena cava detected after peripherally inserted central catheter insertion.

Keywords: Hamburg classification, Truncular lesions, Venogram

CASE REPORT

A 53-year-old African American was sent from Wound Care Clinic to the emergency room because of a positive urine culture for *Klebsiella* done on 06/19/14 at routine check up. The patient denied any fever, chills, nausea, vomiting, diarrhea and body pain. She had an indwelling catheter that was changed two weeks ago as per group home counsellor, and is followed by Wound Care (WC) for a sacral decubitus ulcer. The patient was admitted for Intravenous (IV) antibiotics and infectious disease was consulted with the recommendation of a Peripherally Inserted Central Catheter (PICC) line for the need of two weeks of antibiotics. A left PICC line was placed at bedside without immediate perceived complications. Routine post-procedural chest X-ray revealed the tip of the catheter overlaid the left hilum, not terminating in the superior vena cava, but possibly present within the left superior intercostal vein [Table/Fig-1]. A repeat portable chest X-ray done in the antero-posterior view was obtained. The findings were the same with the tip overlying the left hilum, and not found in the superior vena cava. There was no sign of pneumothorax or pleural effusion. The heart was midly enlarged and the pulmonary vasculature was normal. Impression by the radiologist found it as “no change”. The catheter was not removed and vascular surgery was consulted. The patient consented and brought to the operating room where initial central venogram was carried out through the PICC line.

The left upper extremity and right upper extremity were painted and draped to provide a sterile field. Initial central venogram was carried out through the PICC line which had already been placed. A patent



[Table/Fig-1]: Portable chest X-ray showing the tip of the PICC line overlying the left hilum (red arrow), but not present in the superior vena cava. As illustrated, the lungs are clear without any signs of pathology such as a pleural effusion. **[Table/Fig-2]:** The use of the a glide catheter and guidewire along with simultaneous injections helped confirm the bilateral duplicated superior vena cava (blue arrow) with normal drainage into the right atrium. **[Table/Fig-3]:** Outflow tract showing normal flow from the right ventricle into the right and left pulmonary arteries (blue arrow and lines). (Images from left to right)

left internal juglar, left subclavian, and left innominate with flow along the border of the heart into the right atrium was noted. The decision to confirm a double super vena cava (SVC) was made and the right brachial vein was punctured under ultrasound guidance. A 4 French sheath was inserted. A glide catheter and guidewire were advanced into the right-sided superior vena cava. Catheters were placed in similar positions and simultaneous injections were carried out which confirmed bilateral duplicated superior vena cava with normal confluence in the right atrium and single outflow tract in the pulmonary atery [Table/Fig-2,3]. The PICC line on the the left side was then placed on the left superior vena cava, aspirated, and flushed and capped and secured with occlusive dressing. The right-sided sheath was removed and pressure applied until haemostasis was secured. The simultaneous injection confirmed the diagnosis of persistent left superior vena cava syndrome. The patient tolerated the procedure, was explained the finding and eventually discharged with the PICC line.

DISCUSSION

The human body is comprised of two large venous vessels known as the superior and inferior vena cava in which deoxygenated blood to the right atrium, is “pushed” through the tricuspid valve into the right ventricle, and through the pulmonic valves into the pulmonary artery into the lungs. The deoxygenated blood gets oxygenated while coming into contact with the alveoli of the lungs. The basic physiological mechanism ensures that vital organs such as the brain and other organs are adequalty perfused and are receiving the necessary nutrients to ensure homeostasis in the human body.

Anomalies occuring with respect to the venous circulation are classified into two categories depending on the time when the anomaly occurs in the embryological stage. This classification scheme is defined as the Hamburg classification and allows to differentiate between the two two types of embryological defects – extratruncular or truncular. Defects which occur early on are defined as extratruncular, whereas those defects occuring late in embryogenesis is defined as being truncular. Truncular lesions are more often associated with serious haemodynamic consequences compared to extratruncular lesions due to their direction involvement with the truncal venous system [1]. Persistent Left Superior Vena Cava (PLSVC) is a prime example of truncular venous malformation in addition to other conditions where the pathology of these conditions is due to hypoplastic or hyperplastic vessels/ lesions causing obstruction or dilatation (i.e., internal jugular vein stenosis/aneurysm) depending on the defect [2].

The presence of a persistent left superior vena cava is the most common congenital venous anomaly occurring in the thoracic system. It is present in 0.3% to 0.5% of the general population and up to 10% of patients with a congenital cardiac anomaly [3]. This includes individuals with atrial septal defect, ventricular septal defect, aortic coaractation, transposition of the great vessels, tetralogy of fallot, and anomalous connections of the pulmonary veins [4,5].

The left sided SVC is derived from the left anterior cardinal vein and the left common cardial vein [6]. Embryologically, the cardinal veins are symmetrical and bilateral, however the left cardinal venous system normally obliterates and a new vein drains into the right cardinal vein [3].This anastomosis results in the innominate vein, while the caudal portion regresses to become the “ligament of marshall”. A disturbance during embryology may cause the failure of the normal regression of the left superior cardinal vein, resulting in a persistent left sided vascular structure that empties into the coronary sinus [3]. A persistent left sided SVC is usually asymptomatic, but in up to 8% it can drain into the left atrium increasing the risk of systemic air or particulate emboli from catheter usage (Ghadilai). There are a number of possible drainage systems of the persistent left superior vena cave with 92% through the coronary sinus. In the majority of cases (82-90%) a right sided superior vena cava is also present, or a persistent bridging vein [5]. Other possibilities include the left superior intercostal vein forming a communication between the left superior vena cava and an accessory hemiazygous vein forming a left azygous arch [7].

There are numerous reports of an incidental diagnosis of PLSVC after a central line is noted to take an abnormal left downward course on X- ray [8]. The placement of a central line in patients with PLSVC is possible, however, care must be taken because guide wires, dilators or catheters near the coronary sinus can cause arrhythmias [9].

The management of misplaced catheters depends on their location, indications for central access, and clinical condition of the patient. If there is suspicion that a catheter is misplaced, further workup is indicated prior to removal. If removed, the most significant risk is uncontrolled haemorrhage. Typically the position should be verified with further imaging, such as injection of contrast (liogram/venogram) or cross-sectional CT imaging. It is generally safer to leave the device in situ and consult a vascular surgeon or interventional radiologist rather than a hasty removal with pressure applied to the access site [10,11].

CONCLUSION

This case demonstrates the potential difficulty that may arise when patients with PLSVC require central access. Although no technical problems occurred during placement of the PICC, the patient underwent an additional procedure, which exposed her to radiation along with an increased risk of

vascular injury and acute kidney injury from contrast injection. Familiarity with the common thoracic venous anomalies is helpful in minimising morbidity in patients with PLSVC. PLSVC should be considered whenever a catheter, or guide wire inserted through the left subclavian takes an unusual left sided downward course.

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