

Case Report

Missing inferior vena cava: report of a rare case

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Received: 17 February 2019

Revised: 24 April 2019

Accepted: 30 April 2019

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ABSTRACT

Missing inferior vena cava (IVC) is an uncommon but well recognised anomaly. Our case was a 32 year old male, presented to us with features suggestive of gastric outlet obstruction. When evaluated he was found to have gastric outlet obstruction secondary to cicatrized duodenum and a large subcutaneous vein running from supra pubic region towards the left axilla. Further imaging revealed the complete absence of inferior vena cava and the large collateral draining the venous blood from lower half of the body into left interal jugular vein. Surgical correction of this rare anomaly is not required in the absence of deep vein thrombosis and venous ulcers of lower extremity. Here we report a case of young male with incidentally detected missing IVC as a contribution to the limited literature available on this rare anomaly.

Keywords: Absent IVC, Deep vein thrombosis, Collateral vein

INTRODUCTION

Anomalies of the IVC are well described and recognised less frequently in healthy individuals, but more often in individuals with cardiovascular defects (0.6%-2%).¹ More than 50 distinct inferior vena caval anomalies have been described in the literature.² The anomalies of the IVC includes complete absence, partial absence or presence of bilateral IVC.³ IVC interruption with azygos continuation is a well-recognized anomaly which is usually asymptomatic. Congenital absence of infrarenal IVC with absent deep venous system of the lower extremities is an extremely rare condition that may be associated with significant clinical manifestations.

CASE REPORT

32 year old male, who had past history of two laparotomies, one for duodenal ulcer perforation and bleeding duodenal ulcer, presented to us with complaints

of abdominal pain and recurrent vomiting for 3 months duration. He was evaluated and found to have gastric outlet obstruction secondary to cicatrized duodenum. During examination he was found to have large subcutaneous vein running from supra pubic region towards the left axilla (Figure 1).

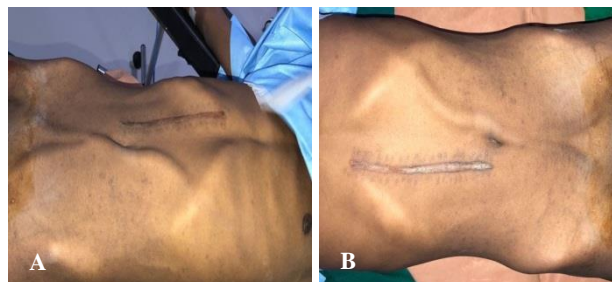


Figure 1 (A and B): Dilated tortuous large subcutaneous collateral vein running from supra pubic region towards the left axilla.



Figure 2: Computed tomography of the abdomen revealing a variant of left internal jugular vein with an external course of vein starting from the inguinal region formed by the union of femoral veins. Inferior vena cava could not be visualized beyond L5 level.

Computed tomography of the abdomen revealed an external course of vein starting from the inguinal region formed by the union of femoral veins, coursing towards left internal jugular vein. IVC could not be visualized beyond L5 level (Figure 2). Since the patient did not have deep vein thrombosis or venous ulcers, the incidentally detected absent IVC was not treated and the patient underwent truncal vagotomy and gastrojejunostomy for the gastric outlet obstruction. The surgery was performed via right paramedian incision without disturbing the variant of left internal jugular vein coursing through the abdominal wall.

DISCUSSION

Absent inferior vena cava (AIVC) is an uncommon but well recognised anomaly. There is controversy as to whether an absent IVC is a true embryonic anomaly or it is the result of peri-natal IVC thrombosis causing gradual regression and eventual disappearance of the IVC.⁴ In normal individuals, IVC is formed by the junction of the common iliac veins anterior to the fifth lumbar vertebral body, slightly to the right of midline. From there it ascends cephalad to receive blood from the renal and hepatic veins on its way. It passes through the aperture in the diaphragm at the level of eighth thoracic vertebra to empty into the right atrium. The azygous vein is a posterior thoracic structure that lies to the right of the spine. This normally receives blood from the right ascending lumbar and lower right intercostal veins. Similarly the hemiazygous vein ascends to the left of the spinal column and receive blood from the left lumbar, the left renal and fourth through seventh intercostal veins. In lower thorax both azygous and hemi-azygous veins course parallel to the descending aorta. At the level of eight thoracic vertebral body the hemi-azygous vein crosses the midline behind the aorta to drain into the azygous vein which in turn empties into the superior vena cava. AIVC is often used to describe three different entities based on the segments of absent IVC (Table 1).⁵

Absence of the intrahepatic segment of IVC with azygous continuation occurs in 0.6% of patients with congenital heart disease. These include situs inversus, asplenia, or polysplenia, persistent left superior vena cava and congenital pulmonary venolobar syndrome. More recently, isolated absent IVC has been recognized as a solitary, incidental finding in asymptomatic patients.⁶

Table 1: Types of absent IVC.

Type	Basic pathology	Associated conditions
Absence of suprarenal IVC	Failure to form the right subcardinal vein	Associated cardiac and visceral anomalies
Absence of infrarenal IVC	Failure of development of right supracardinal vein	Preservation of suprarenal segment
Absence of entire IVC	All three paired vein systems failed to develop	Nil congenital anomalies

Gayer et al recommended that all patients with an IVC anomaly be screened for the presence of thrombophilic disorder.⁷ It has been suggested that this finding could represent an acquired defect caused by perinatal renal vein thrombosis although most researchers believe embryologic dysgenesis is the cause. It is hypothesized that blood return with an absent IVC is inadequate, despite adequate collaterals, resulting in chronic venous hypertension in the lower extremities causing venous stasis which precipitates thrombosis. Multiple emboli from DVT in the common and superficial femoral veins migrate through the well developed hemi-azygous and/or azygous system to the pulmonary circulation.

Idiopathic DVT is one of the manifestations of missing IVC. Absence of the IVC cannot be detected by ultrasonography, and so a CT scan was recommended in all young patients with an idiopathic DVT. The most reliable non-invasive methods for diagnosing IVC anomalies are CT with intravenous contrast or Magnetic Resonance scan. CT scan, unlike US, is a good imaging modality of the retro-peritoneal space.⁸ Another accurate, but more invasive, imaging modality is venography, which is particularly useful if any surgery is planned.

AIVC may be diagnosed incidentally on chest radiography, which will often show a paraspinous shadow adjoining the right hemidiaphragm. There has been an increased recognition of this anomaly with the more prevalent use of computed tomography for other indications, which off late has become the single best study to diagnose AIVC. This carries the advantage of detecting other associated anomalies. Color flow duplex scan of the lower limb can help exclude venous obstruction or venous incompetence. If duplex is suggestive, then there is a role for ascending venography.

There is very little evidence available on the surgical correction or treatment of this uncommon anomaly. Surgical repair with prosthetic graft from the iliac vein to the intra-thoracic azygous vein has been reported in a case of complete absence of the IVC but patent iliac veins and non-healing pre-tibial venous ulceration. The ulcer healed within thirty days of IVC repair.⁹ However, since Knudtzon described in 1986 a 13 year old boy with absence of the entire inferior vena cava that was diagnosed by venography, computer tomography and ultrasonography after he was found to have dilated superficial abdominal veins since birth. He was found to have an intact extensive collateral circulation from the left femoral vein, right common iliac vein and the kidneys, to the azygos vein and superior vena cava via the deep abdominal veins. He stated that surgical treatment was neither necessary nor advisable in asymptomatic absent IVC.¹⁰ This statement holds good even after three decades.

CONCLUSION

In the absence of past history of idiopathic varicose ulceration with evidence of chronic venous hypertension in young adults, incidentally detected missing IVC can be observed. The very limited data from the literature suggests extensive evaluation to exclude an intra-abdominal deep venous anomaly and non surgical management in the absence of one. However there is no consensus regarding the use of anti-coagulation given the possible risk of further DVT and pulmonary embolism. Surgery is not mandatory, but extensive search for the associated anomalies such as renal atrophy or agenesis can themselves be of significant clinical importance.

Funding: No funding sources

Conflict of interest: None declared

Ethical approval: Not required

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Cite this article as: Perumal SK, Subramaniam SRV. Missing inferior vena cava: report of a rare case. Int Surg J 2019;6:2201-3.