Agnesis of Inferior Vena Cava with Cavernous Transformation and Hypoplastic Infrarenal Part of IVC, Rare Congenital Vascular Anomalies: A Case Series

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Abstract: Congenital vascular anomalies are although rare but may cause serious complications. Vascular pathologies are best demonstrated by contrast enhanced computed tomography, with the advent of multidetector computed tomography and applying multiplanar reformation techniques. However, ultrasound color Doppler and catheter angiography are accessory imaging modalities utilized to rule out vascular anomalies. We present here a case series of 2 patients aged 19 years and 54 years, diagnosed with congenital vascular anomalies (agenesis of inferior vena cava and hypo plastic infra renal segment of inferior vena cava). The development of inferior vena cava (IVC) includes early embryogenesis stages including formation, confluence and involution of three primary longitudinal venous systems. Agenesis of inferior vena cava or hypoplasia of segment of inferior vena cava leads to formation of collaterals to maintain the venous flow. Contrast enhanced computed tomography with multiplanar reformation provides ulterior spatial and contrast resolution in cases of recognized vascular abnormalities. Inferior vena cava obstruction due to thrombosis or congenital vascular anomaly leads to extensive collateralization accompanied by varicose veins. In our case series, we found contrast enhanced computed tomography as an ulterior modality in setting ultimate diagnosis with accuracy and maximum detail.

Key words: IVC, hypoplastic, azygous vein, cavernous transformation, varicose.

1. Introduction

Anomalies of inferior vena cava are present in 0.3%-0.5% of otherwise healthy individuals and in 0.6%-2% of patients with other cardiovascular defects [1, 2]. The prevalence of deep venous thrombosis (DVT) varies according to age being 10 times lower in 20-40 years old individuals than in older age groups [3, 4]. In young patients, its etiology is frequently associated with risk factors such as congenital and acquired thrombophilia, autoimmune diseases, pregnancy and puerperium, use of oral contraceptives, neoplasms, surgical procedures, prolonged immobilization, and trauma [5-7]. In adults, absent inferior vena cava (AIVC) can cause diagnostic problems in the paravertebral regions because of their tumor like appearance [8, 9].

2. Materials and Methodology

This case series was held at Jinnah Postgraduate Medical center Karachi, Pakistan. Both the patients were referred from out patients department (OPD) for computed tomography (CT). Axial post contrast (Iopamiro 370) imaging was performed for abdomen pelvis, with 2 mm slice thickness, with an interspace of 2 mm, 120 KVp, 300 mA, axial images were reconstructed into coronal and sagittal planes.

3. Case Report 1

A 19 year old male, who came to outpatient department (OPD) with complain of lower limb varicose veins, and generalized abdominal pain. Patient was sent to radiology department for abdominal imaging. Abdominal ultrasound, a venous duplex scan of both legs was performed. On duplex both legs were

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edematous with varicosities without any evidence of deep venous thrombosis and deep or superficial venous incompetence. On abdominal sonogram, we revealed dilated anterior abdominal wall veins while no hepatomegalia/splenomegaly, nor portal hypertension was observed. A post contrast CT abdomen was suggested following abdominal sonography, CT scan revealed absent inferior vena cava (AIVC) (Fig. 1) with cavernous transformation, enlarged azygous and hemiazygous veins with a cluster of collaterals in the pelvic region as well as anterior abdominal wall (Fig. 2). CT scan revealed no venous thrombosis.

Patient was suggested to avoid prolonged immobilization maintain lower limb elevation while taking on bed, as those patients with agenesis of IVC were found to be at a greater risk of developing deep venous thrombosis.

4. Case Report 2

A 54 year old female came to outpatient department (OPD) complaining of lower limb pain was sent to radiology department for CT scan abdomen post contrast study. Post contrast axial images were obtained and were reformatted into coronal and sagittal

Fig. 1 Post contrast computed tomography MPR coronal image showing complete absence of inferior vena cava.

(A)

(B)

(C)

Fig. 2 Post contrast axial Computed Tomography images showing (A) dilated azygos and hemiazygos, (B) extensive patent anterior abdominal wall collaterals and dilated paravertebral veins and (C) collateral vessels in the pelvic region.

planes. Findings were, absent/hypoplastic infra renal part of IVC not showing any contrast enhancement associated with dilated gonadal veins (Fig. 3) on both

sides, predominantly on left side throughout its course. Lumbar and azygous veins were also prominent consistent with collateralization (Fig. 3).

5. Discussion

The IVC is developed as a result of a complex embryologic process between the sixth and eighth weeks of gestation. Three pairs of primitive veins (postcardinal, subcardinal, and supracardinal) appear in this order and give rise to the four segments of the adult IVC: hepatic, suprarenal, renal, and infrarenal (Fig. 4)

[3, 4, 9, 10]. AIVC is often used to describe three different entities:

(1) Absence of the suprarenal IVC results from failure to form the right subcardinal vein. The hepatic segment drains directly into the right atrium, and the blood from the infrarenal IVC returns to the heart through the azygos and hemiazygos veins [8, 10]. There is association with other cardiac and visceral anomalies, such as dextrocardia, atrial septal defect, atrioventricular canal, situs inversus, polysplenia, or asplenia [9, 11, 12].

Fig. 3 Contrast Enhanced CT shows absent infra renal part of IVC with prominent azygous and hemi azygous vein and dilated bilateral gonadal veins.

Fig. 4 Embryologic deviation of the IVC from 7 weeks of gestation (A) to the adult (B).

(2) Absence of the infrarenal IVC with preservation of the suprarenal segment implies a failure of the development of the right supracardinal vein [10].

(3) Absence of the entire IVC, as in our patient's case, suggests that all three paired vein systems failed to develop properly [10], but it has no relation to the other congenital anomalies described previously [11].

The reasons for the developmental failure are unclear. One hypothesis is embryonic dysontogenesis [1, 11, 13], but some authors suggest that it is the result of an intrauterine or perinatal thrombosis [9]. AIVC could be present with DVT. An inadequate blood return through collaterals may increase the venous blood pressure in the veins of the legs, thus facilitating deep venous thrombosis (DVT) [9, 13]. The dysgenesis of the IVC has been described in coincidence with clotting defects [1, 11, 13, 14]. This paper describes a case of complex congenital malformation with absent IVC in association with azygous continuation, patent anterior abdominal wall venous collaterals, varicocoele, and lower limb varicose veins.

Normally, the IVC is formed by the junction of the common iliac veins anterior to the fifth lumbar vertebral body, a little to its right [15]. It ascends cephalad to receive blood from the renal and hepatic veins and having passed through the diaphragm to empty into the right atrium. The azygous vein, a posterior thoracic structure that lies to the right of the spine and empties into the superior vena cava, 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 receives blood from the left lumbar, the left renal and fourth through seventh intercostal veins [16]. In lower thorax, both azygous and hemi-azygous veins course parallel to the descending aorta. At the level of eighth thoracic vertebral body, the hemi-azygous vein crosses the midline behind the aorta to drain into the azygous vein [15]. The reasons for the developmental failure are unclear. One hypothesis is embryonic dysontogenesis [1, 11, 12], but some authors suggest

that it is the result of an intrauterine or perinatal thrombosis [10]. But in our case, patient did not presented with DVT despite of dysgenesis of IVC.

6. Conclusion

In our case series, we found that congenital IVC abnormalities may not present clinically for a long time due to collateral function. Peripheral venous thrombosis or varicose veins are often first symptoms of congenital anomaly of IVC. So in case of DVT or varicose veins, a potential cause of IVC abnormality must always be excluded with the help of contrast enhanced computed tomography.

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