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Case Report

Congenital infra renal agenesis of inferior vena cava - an uncommon entity in clinical practice

Saranya Ayyadurai*, Rajoo Ramachandran, Haree Shankar M, Venkatasai P. M.

Department of Radiology and Imaging Sciences, Sri Ramachandra Medical College, Chennai, India

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***Correspondence:**
Dr. Saranya Ayyadurai,
E-mail: saranya_thiru@yahoo.com

ABSTRACT

Agenesis of the inferior vena cava is an uncommon congenital anomaly and can be misdiagnosed if unaware. This condition is one of the important risk factors for the development and recurrence of deep vein thrombosis in young people. In this case report we discuss the imaging findings of this uncommon entity on computed tomography in a young female who presented with lower leg pain post-delivery. This vascular abnormality is mostly incidentally diagnosed in adults and only fewer cases have been reported in English literature.

Keywords: Congenital agenesis, IVC, Contrast enhanced CT

INTRODUCTION

Embryogenesis of IVC is a complex process involving the formation and anastomoses between three pairs of embryonic veins. Persistence or regression of these embryonic veins can lead to numerous variations that can result in anomalies such as complete agenesis, isolated left IVC, double IVC, and other variants.

Incidence of agenesis of inferior vena cava in the general population is 0.3 - 0.5%.[1,2] The mean age of onset of clinical symptoms is between 2nd and 4th decade with slight male predominance.[3]

CASE REPORT

A 29-year-old female immediate postpartum presented with lower abdominal pain. Baseline investigations were done and found normal. Patient was referred for CECT abdomen, which showed complete absence of infra renal IVC and common iliac veins and its branches (Figure 1a and 1b). The presacral veins, ascending lumbar veins and para vertebral veins joins to form a collateral plexus of renal veins (Figure 2) which drain into the left renal vein and further draining into left sided IVC. The left renal and right renal vein unites to form a normal right supra renal IVC (Figure 3). Patient was screened for lower limb DVT and advised for regular follow up.

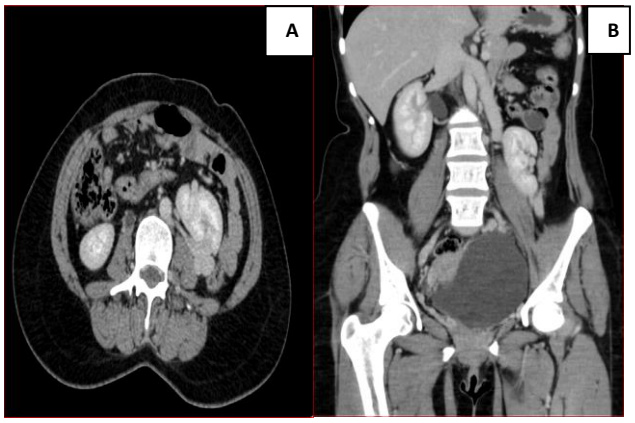

Figure 1 A and B: Complete absence of infra renal IVC, common iliac veins and its branches.

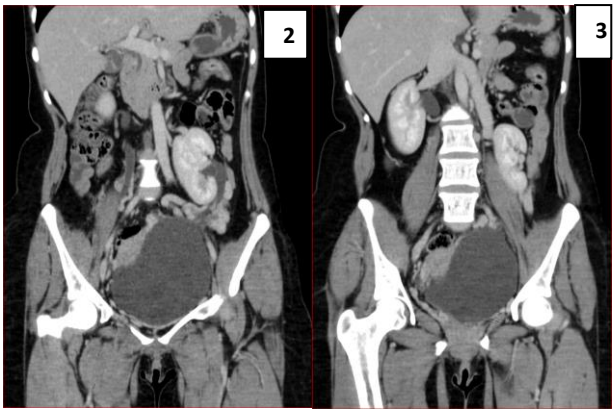

Figure 2 and 3: The presacral veins, ascending lumbar veins and para vertebral veins joins to form a collateral plexus of renal veins which drain into the left renal vein and further draining into left sided IVC.

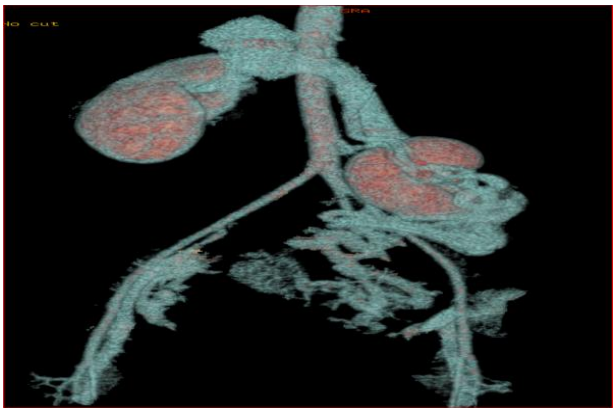

Figure 4: Volume rendered imaging shows complete absence of infrarenal IVC with multiple collaterals forming left renal vein which joins with right renal vein to form supra renal IVC.

DISCUSSION

Normal adult right-sided inferior vena cava (IVC) is complete by the 8th week of gestation. The development of the IVC is a complex process involving the formation of several anastomoses between three paired embryonic veins: the posterior cardinal veins, the sub cardinal veins, and the supracardinal veins, which arise in a chronological order between 4 to 8 weeks of fetal life. The infrahepatic IVC develops from these three pairs of veins through the processes of fusion, regression, and formation of midline anastomoses between them.[4]

The final adult form of the IVC is formed of four segments: the hepatic segment formed from the vitelline vein, suprarenal segment from the sub cardinal hepatic vein anastomosis, renal segment from the right supra-cardinal and sub cardinal vein anastomosis and infrarenal segment from the right supracardinal vein.

The common congenital anomalies of IVC are isolated left IVC, double IVC, agenesis of a segment or total IVC, azygous and hemiazygous continuation of IVC.

An isolated left IVC occurs in 0.2-0.5% of the population. This anomaly results from the regression of the right supra Cardinal vein and persistence of the left supra cardinal vein. The left IVC ascends and joins the left renal vein.[5]

A double IVC occurs in 1-3% of the population. It results from persistence of both the left and right supra- cardinal veins. Usually, the left IVC ends at the level of the left renal vein, crossing over to join the right-sided IVC. There may be variations in the size of the left and right IVC.

Atresia or agenesis is due to absence of fusion of one or multiple segments of IVC. Azygous continuation of IVC occurs in the absence of hepatic segment of IVC.[4] Our patient was diagnosed to have complete infra renal agenesis of IVC with absence of common iliac veins and its branches. Some studies say that complete absence of IVC is due to the prenatal or infrauterine thrombosis.

Patients usually present with acute abdominal pain, right lower extremity pain, inflammation, erythema, decreased mobility, stiffness and pain in right leg following physical activity, rarely patients come with pulmonary thromboembolism.

On ultrasound IVC agenesis appear as lack of continuity of the IVC segment and the presence of collateral venous circulation in the abdominal wall or retroperitoneal location.[6] Thrombus in the lower limb venous system can be assessed with color Doppler. Contrast enhanced CT and MR are superior in assessment of the IVC anomalies over USG.[7]

Deep vein thrombosis (DVT), especially in young adults is the common complication of IVC agenesis.[8] This may be due to slow flow in the prominent collateral vessels, which are much lesser in caliber than normal IVC. Chronic venous stasis, impaired venous return, and venous hypertension due to absent main venous pathways and associated thrombophilia may be contributing factors. DVT is bilateral in more than 50% of patients.[9]

Anticoagulants and elastic stockings help in the symptomatic management of IVC agenesis. Patients should be advised to avoid other risks of thrombosis like immobilization and oral contraceptive [10]. Few have required surgical intervention for relief of symptoms. Follow up studies on these patients determine their recurrence, length of treatment, alternate treatment options, and prognosis.

CONCLUSION

Congenital anomalies of the IVC result from its complex embryological development. Young adults with deep vein

thrombosis should be suspected for IVC agenesis. As IVC agenesis could not be corrected, lifetime precautions should be taken to avoid DVT.

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