Absence of Infrarenal Portion of the Inferior Vena Cava With Acute Lower Extremities Venous Thrombosis: A Case Report

Susan Mohammadi¹, Nastaran Hesam Shariati², Fardin Fathi³, Siamak Arshadi⁴, Fahimeh Rajabi⁴, Mohammad Bakhtiar Hesam Shariati⁵

¹ Department of Radiology, Faculty of Medicine, Kurdistan University of Medical Sciences, Sanandaj, Iran

² School of Medical Sciences, Faculty of Medicine and Health, University of Sydney, Camperdown, Australia

³ Cellular and Molecular Research Center, Research Institute for Health Development, Kurdistan University of Medical Sciences, Sanandaj, Iran

⁴ Department of Radiology, Besat Hospital, Kurdistan University of Medical Sciences, Sanandaj, Iran

⁵ Department of Anatomical Science, Faculty of Medicine, Kurdistan University of Medical Sciences, Sanandaj, Iran

Received: 28 Apr. 2020; Accepted: 12 Nov. 2020

Abstract- A lack of congenital Inferior Vena Cava (IVC) is an uncommon malformation that has been identified in combination with idiopathic Deep Venous Thrombosis (DVT), exclusively. It may not even be revealed during the lifetime. A 63-year-old female was accepted with three months of abdominal and pelvic pain and localized edema on the right flank. During this admission, she was examined and recognized for deep vein thrombosis (DVT). Ct scan images showed a lack of the Inferior Vena Cava with enormous thrombosis collaterals of the superficial vein in the abdomen. In this case report, we report a woman with side pain who has an absence of the IVC.

 $\ensuremath{\mathbb{O}}$ 2020 Tehran University of Medical Sciences. All rights reserved.

Acta Med Iran 2020;58(12):658-661.

Keywords: Congenital abnormalities; Vena cava; Inferior; Venous thrombosis

Introduction

Malformations of the venous system are not rare. Approximately 60 distinguished inferior vena cava malformations have been observed in the literature (1). Absence of inferior vena cava (AIVC) Continuing with azygos is a well-known malformation that can be observed in asymptomatic patients (1). Nevertheless, lack of congenital of infrarenal IVC along with a lack of a deep venous system of the lower limb is a very unusual situation that may be accompanied with considerable clinical advents (2).

A malformation of the inferior vena cava (IVC) is in 0.3% to 0.5% of otherwise healthy people 1-3 and in 0.6% to 2% of patients is accompanied by other cardiovascular Complications (3-6). Lack of the IVC has an occurrence of 0.0005% to 1% in the public population, eight and last reports acknowledge its role as a powerful predisposing operating for the growth of deep venous thrombosis (DVT) in young people (7,8). Deep venous thrombosis (DVT) is related due to its high frequency and illness/fatality rates. The prevalence of this disorder in the western people is appraised to be

1:1,000 Persons per year (9,10). The incidence rate changes by age, And in people 20 to 40-year-old, it is ten times lower than in the older age groups. The Increasing use of medical imaging devices has made it possible for physicians to recognize most of the time the presence of inferior vena cava (IVC) anomalies along with DVT of the lower limbs (11,12). Here, we present a case of a late presentation of DVT with an underlying etiology of a Lack of IVC.

Case Report

A 65-year-old female presented to Bastat Hospital of Sanandaj with abdominal pains and Inflation over the right flank for a Multi-month course. Recently, she noticed severe swelling in her right thigh. She was a non-smoker, and there was no considerable family history of the disease. She had no Digestive and cardio-respiratory problems and diseases. There were no significant changes in appetite and body weight. She had a history of two pregnancies with cesarean section and a family history of Intravenous varices in the lower limbs. Blood test results were normal. Ultrasound showed

clearly that the Common and superficial femoral, external iliac, proximal part of the saphenous vein on the left and right had venous thrombosis. Computed tomography abdomen to pelvis did not show any malignancy and visceral problems, but the vascular findings and studies in CT scan were consistent with ultrasound. CT scan images showed clearly that the bilateral iliac veins are not seen, and the pelvic veins, especially on the left, were dilated and tortuous. Also, bilateral femoral veins drain into the Pelvic vessels. CT images confirm and indicate that the patient has a congenital absence of the IVC malformations (Figure 1 & 3). Also, a dilated vein is drained from the pelvic area to the left renal vein, and probably this is the left ovarian vein that has acute thrombosis (Figure 1). Also, CT images showed clearly that the Common femoral vein on the left and right had venous thrombosis (Figure 2 and 3).

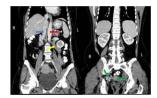


Figure 1. Coronal CT-scan images with IV and oral contrast of the abdominal and pelvic region. Reveal the absence of inferior vena cava with acute venous thrombosis of lower extremities. The left ovarian vein (yellow arrow), Suprarenal portion of inferior vena cava (blue arrow), Pelvic veins (green arrow), and Aortic artery (red arrow)

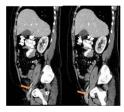


Figure 2. Sagittal CT-scan images with IV and oral contrast of the abdominal and pelvic region reveal the absence of inferior vena cava with acute venous thrombosis of lower extremities. The right femoral

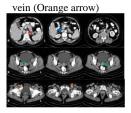


Figure 3. Axial CT-scan images with IV and oral contrast of the abdominal and pelvic region reveal the absence of inferior vena cava with acute venous thrombosis of lower extremities. The left ovarian vein (yellow arrow), Suprarenal portion of inferior vena cava (blue arrow), Pelvic veins (green arrow), femoral vein (Orange arrow), and aortic artery (red arrow)

Discussion

The spectrum of IVC congenital malformations is well defined (13,14). Congenital types of the infrarenal IVC are believed to have an outbreak of less than 2% in the normal society, with complete lack of the IVC happening in 0.3% of healthy individuals and Patients (15-17). CT scan has been suggested as the procedure of choice for detecting the presence of hemi-azygous and azygous system as main clues to, Specific types of IVC anatomy (17).

AIVC is mostly applied for describing three different existences: 1. a lack of the suprarenal IVC results of the defect information the right sub cardinal vein. The hepatic part is poured directly inside the right atrium, and blood returns to the heart from infrarenal IVC through the azygos and hemiazygos veins (8,14). This type of abnormality of the absence of inferior vena cava (AIVC) is associated with a number of abnormalities, dextrocardia, atrial including septal defect atrioventricular canal (18,19). 2. A lack of the infrarenal IVC, as in our patient's case, by the preservation of the suprarenal part, refers to a Defect in the growth of the right supra-cardinal vein (14). 3. A lack of the whole IVC shows that all three paired vein systems have not grown properly (17) And it has nothing to do with the congenital malformations already described (19).

The causes of IVC in writing are controversial. Some researchers believe that thrombosis on the IVC in the perinatal course is the source of its disappearance. Therefore no embryologic malformations are observed (20). Lack of IVC formation may be associated with other congenital malformations like sphenic anomalies, renal agenesis, dextrocardia and etc. (21,22). These malformations are specified in over 1% of people, but the occurrence of maybe created on 2% of the people who have congenital heart disease (23). In complete lack of IVC, the veins drain their blood via pelvic and abdominal, thoracolumbar veins, which may result in the Signs in the thorax, lumbar and genital parts before creating of lower limbs' DVT (4). However, these signs are uncommon and indeterminate; their early diagnosis in young patients may show the existence of IVC anomalies (12). Patients with IVC abnormalities are at risk for DVT due to lower limb venous arrest (22). In the case described, the patient had iliofemoral, and the proximal part of the large saphenous vein DVT of the left lower limb, which could be related to a significant reduction left common iliac vein Viewed on ct image.

So this malformation shows almost 5% of deep vein thrombosis (DVT) in people <30 years of age (24,25).

Absence of infrarenal portion of the inferior vena cava

The IVC during pregnancy is caused by a complex reaction involving posterior cardiac anastomosis. The IVC is caused in the embryonic period by a series of complex reactions resulting from the anastomosis of the posterior cardinal, sub cardinal, and supra-cardinal veins to form a One-way system on the right. AIVC is one type of vascular malformation that results from the defeat of these paired combinations to fuse. The collateral and parallel deep venous system that creates are likely insufficient, and it will not be able to counteract increased blood flow. This inability to cope with hypertension leads to chronic venous hypertension in the lower extremities with stasis in the veins, and this can lead to thrombosis in the veins (24).

The ideal medical imaging device for identification of an IVC disorder must have high Power of detection and be safe and able to rebuild images. It is difficult to recognize any IVC malformation by the medical ultrasound machine.

There are several clues to radiological imaging that can help detect the absence of IVC or abnormalities (26). One of the most common, useful, and non-invasive methods to detect these abnormalities are CT scan with contrast of the vessels or Magnetic Resonance Imaging (27). The most appropriate treatment for these disorders is using anticoagulant therapy for at least six months. Also, if the patient discontinues the drug before this period, the risk of recurrence is high (22).

The absence of Inferior Vena Cava (AIVC) is an uncommon malformation that may not even be revealed during the lifetime. Nevertheless, AIVC, along with a lack of a deep venous system of the lower limb, is a very unusual situation that may be accompanied by considerable clinical advents.

Today, computerized tomography (CT) scan and Magnetic resonance imaging (MRI) has revolutionized the diagnosis of diseases and lesions (28).

Ethical Approval and Consent to Participate

This research has been confirmed by the Research Center of Kurdistan University of Medical Sciences and Ethics Committee with the file number IR.MUK.REC.1399.084

Consent for Publication

Written informed consent was obtained from a legally authorized representative(s) for anonymized patient information to be published in this article which was approved by the Research Center of Kurdistan

University of Medical Sciences.

Competing Interests

All authors declare that there is no conflict of interest that prejudices the impartiality of this scientific work.

Acknowledgments

The authors are grateful to all the staff of Beast Hospital in Sanandaj.

References

- Spentzouris G, Zandian A, Cesmebasi A, Kinsella CR, Muhleman M, Mirzayan N, et al. The clinical anatomy of the inferior vena cava: a review of common congenital anomalies and considerations for clinicians. Clin Anat 2014;27:1234-43.
- Gupta A, Kumar S, Kothari SS. Congenital absence of infrarenal inferior vena cava and deep veins of the lower limbs: a case report. J Med Case Rep 2016;10:218.
- Gil RJ, Pérez AM, Arias JB, Pascual FB, Romero ES. Agenesis of the inferior vena cava associated with lower extremities and pelvic venous thrombosis. J Vasc Surg 2006;44:1114-6.
- 4. Gayer G, Luboshitz J, Hertz M, Zissin R, Thaler M, Lubetsky A, et al. Congenital anomalies of the inferior vena cava revealed on CT in patients with deep vein thrombosis. AJR Am J Roentgenol 2003;180:729-32.
- Vijayvergiya R, Bhat M, Kumar R, Vivekanand S, Grover A. Azygos continuation of interrupted inferior vena cava in association with sick sinus syndrome. Heart 2005;91:e26.
- 6. Ramanathan T, Michael T, Hughes D, Richardson AJ. Perinatal inferior vena cava thrombosis and absence of the infrarenal inferior vena cava. Journal of vascular surgery. 2001;33(5):1097-9.
- Darrel S, Isam H, Armando S. Absence of the retrohepatic inferior vena cava: what the surgeon should know. Am Surg 2005;71:502-4.
- 8. Vermeulen E, van Urk H. Agenesis of the inferior vena cava. Eur J Vasc Endovasc Surg 1996;12:493-6.
- Cho BC, Choi HJ, Kim SK, Lee YW, Lee DH, Hong YK, et al. Congenital absence of inferior vena cava as a rare cause of pulmonary thromboembolism. Yonsei Med J 2004;45:947-51.
- Iqbal J, Nagaraju E. Congenital absence of inferior vena cava and thrombosis: a case report. J Med Case Rep 2008:2:46.
- 11. Suh HJ, Kim WT, Kim MY, Cho YK. Combined

- anomaly of the right hepatic lobe agenesis and absence of the inferior vena cava: a case report. Korean J Radiol 2008;9:S61-S4.
- 12. García-Fuster MJ, Forner MJ, Flor-Lorente B, Soler J, Campos S. Inferior vena cava malformations and deep venous thrombosis. Rev Esp Cardiol 2006;59:171-5.
- 13. Alicioglu B, Kaplan M, Ege T. Absence of infrarenal inferior vena cava is not a congenital abnormality. Bratisl Lek Listy 2009;110:304-6.
- 14. Bass JE, Redwine MD, Kramer LA, Huynh PT, Harris Jr JH. Spectrum of Congenital Anomalies of the Inferior Vena Cava: Cross-sectional Imaging Findings 1: (CME available in print version and on RSNA Link). Radiographics 2000;20:639-52.
- 15. Bapat G, Daly K, Razzaq R, Onwudike M. Unusual presentations of congenital absence of the inferior vena cava. Int J Angiol. 2003;12:48-50.
- 16. Zhou W, Rosenberg W, Lumsden A, Li J. Successful surgical management of pelvic congestion and lower extremity swelling owing to absence of infrarenal inferior vena cava. Vascular 2005;13:358-61.
- 17. Cooper M, Waldo O, Davis B, Duerinckx AJ. Absent infrarenal inferior vena cava. Radiol Case Rep 2011;6:535.
- 18. Dougherty MJ, Calligaro KD, DeLaurentis DA. Congenitally absent inferior vena cava presenting in adulthood with venous stasis and ulceration: a surgically treated case. J Vasc Surg 1996;23:141-6.
- 19. Yun SS, Kim JI, Kim KH, Sung GY, Kim JS, Moon IS, et al. Deep venous thrombosis caused by congenital absence inferior vena cava, combined hyperhomocysteinemia. Ann Vasc Surg 2004;18:124-9.

- 20. Bass J, Redwine M, Kramer L, Harris Jr J. Absence of the infrarenal inferior vena cava with preservation of the suprarenal segment as revealed by CT and MR venography. AJR Am J Roentgenol 1999;172:1610-2.
- 21. Atmatzidis K, Papaziogas B, Pavlidis T, Paraskevas G, Mirelis C, Papaziogas T. Surgical images: soft tissue: reccurent deep vein thrombosis caused by hypoplasia of the inferior vena cava. Can J Surg 2006;49:285.
- 22. Konopka CL, Salame M, Padulla GA, Muradás RR, Batistella JC. Agenesis of inferior vena cava associated with deep venous thrombosis. J Vasc Bras 2010;9:196-9.
- 23. Felicio ML, Martins AS, Andrade RRd, Silva MAdM. Ausência parcial de veia cava inferior associada à malformação intestinal. Braz J Cardiovasc Surg 2007;22:362-4.
- 24. Lambert M, Marboeuf P, Midulla M, Trillot N, Beregi J-P, Mounier-Vehier C, et al. Inferior vena cava agenesis and deep vein thrombosis: 10 patients and review of the literature. Vasc Med 2010;15:451-9.
- 25. Obernosterer A, Aschauer M, Schnedl W, Lipp RW. Anomalies of the inferior vena cava in patients with iliac venous thrombosis. Ann Intern Med 2002;136:37-41.
- 26. Koc Z, Oguzkurt L. Interruption or congenital stenosis of the inferior vena cava: prevalence, imaging, and clinical findings. Eur J Radiol 2007;62:257-66.
- 27. Ueda J, Hara K, Kobayashi Y, Ohue S, Uchida H. Anomaly of the inferior vena cava observed by CT. Comput Radiol 1983;7:145-54.
- 28. Farashah MSG, Shariati NH, Asghari R, Mohamadi S, Dolatkhah MA, Shariati MBH. Pulmonary Sequestration: A Case Report. Acta Med Iran 2019;57:398-401.