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Where is the Inferior Vena Cava? A Case Report

Gdje je *vena cava inferiror*? – prikaz slučaja

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Abstract. ***Aim:*** The absence of the inferior vena cava (IVC) is a rare abnormality reported in less than 1% of the population. The collateral veins are enlarged with agenesis IVC, enabling blood transport to the superior vena cava. IVC agenesis (IVCA) may predispose to venous hypertension and complications, including thromboembolism. ***Case report:*** We present a case of a 20-year-old female patient with syncope who came to the emergency ambulance. She had profuse perspiration, and she lost 12 kg of body weight in one month. Under the suspicion of pulmonary thromboembolism, a CT (computed tomography) angiography of the pulmonary arteries showed a retrosternal para-aortic hypovascular soft tissue formation with a diameter of 30×40×85 mm. Laboratory findings also confirmed higher thyroid hormone values. Immunohyperthyroidism was diagnosed. A venous phase of CT angiograpy scan of the thorax was performed which showed that para-aortic hypovascular soft-active formation is the thymus, but an incidental finding was also discovered (Figure 1). There was no representation of the inferior v. cava in the area from the renal veins to the heart, and the dominant azygos and hemiazygos v. were visible (Figure 2). Hemiazygos vein merges into the azygos vein in the thorax area (Figure 3). All the above corresponds primarily to the anatomical variant of congenital absence of v. cava inferior. ***Conclusion*****:** IVCA is a significant but highly under recognized cause of unprovoked DVT (deep vein thrombosis) of the lower extremities in the young population without additional risk factors. Therefore, in unclear cases of unprovoked deep vein thrombosis of the lower extremities in the younger population, this cause should also be considered, and accordingly, the necessary diagnostic evaluation should be performed, which includes imaging for vascular anomalies along with a thrombophilic screen. Treatment is mainly conservative and may require lifelong anticoagulation therapy.

Keywords: computed tomography angiography; inferior vena cava; thromboembolism

Sažetak. ***Cilj:*** Izostanak donje šuplje vene (engl. *inferior vena cava*; IVC) rijetka je abnormalnost koja se javlja u manje od 1 % populacije. Kolateralne su vene proširene kod ageneze IVC-a, omogućujući prijenos krvi u gornju šuplju venu. Ageneza IVC-a (engl. *inferior vena cava agenesis*; IVCA) može predisponirati venskoj hipertenziji i komplikacijama, uključujući tromboembolizam. ***Prikaz slučaja:*** Predstavljamo slučaj 20-godišnje pacijentice koja je došla u hitnu ambulantu zbog sinkope. Imala je obilno znojenje i izgubila je 12 kilograma tjelesne težine u razdoblju od mjesec dana. Zbog sumnje na plućnu tromboemboliju, učinjena je CT angiografija (engl. *computed tomography*) plućnih arterija koja je pokazala retrosternalnu paraaortnu hipovaskularnu formaciju mekih tkiva promjera 30 × 40 × 85 mm. Laboratorijski nalazi također su potvrdili povišene vrijednosti hormona štitnjače. Dijagnosticiran je imunohipertireoidizam. Učinjena je venska faza CT angiografije toraksa koja je pokazala da se paraaortna hipovaskularna formacija mekih tkiva odnosi na timus, ali je pritom otkriven i slučajan nalaz (Slika 1). Nije bilo prikaza donje šuplje vene (*vena cava inferior*) u području od bubrežnih vena do srca, dok su dominantno bile prikazane vene azigos i hemiazigos (Slika 2). Vena hemiazigos ulijeva se u venu azigos u području toraksa (Slika 3). Sve navedeno primarno odgovara anatomskoj varijanti kongenitalnog izostanka donje šuplje vene. ***Zaključak:*** Ageneza donje šuplje vene značajan je, ali izrazito nedovoljno prepoznat uzrok neprovocirane duboke venske tromboze (engl. *deep vein thrombosis*; DVT) donjih ekstremiteta kod mlađe populacije bez dodatnih rizičnih čimbenika. Stoga, u nejasnim slučajevima neprovocirane duboke venske tromboze donjih ekstremiteta kod mlađih osoba, treba uzeti u

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obzir i ovu mogućnost te prema tome provesti odgovarajuću dijagnostičku obradu koja uključuje slikovne metode za otkrivanje vaskularnih anomalija uz trombofilijski probir. Liječenje je uglavnom konzervativno i može biti potrebna doživotna antikoagulantna terapija.

Ključne riječi: donja šuplja vena; računalna tomografska angiografija; tromboembolizam

Congenital absence of the inferior vena cava (IVCA) is a rare vascular anomaly. IVCA is a significant but highly under-recognized cause of unprovoked DVT of the lower extremities in the young population without additional risk factors.

INTRODUCTION

The etiology of infrarenal IVC absence is controversial. An absent infrarenal inferior vena cava can be congenital, due to the failure of development of the posterior cardinal and supracardinal veins. However, this does not explain the failure of the postcardinal veins to persist. A single embryological event does not fully explain infrarenal IVC absence. On the other hand, it has been also suggested that infrarenal absence of the IVC is not of embryonic origin but the result of intrauterine or perinatal thrombosis of IVC. It is an extremely rare anomaly[1].

The patient may present with symptoms of lower extremity venous insufficiency or idiopathic deep venous thrombosis, particularly in younger patients.

The non-invasive modalities of choice are contrast enhanced CT or MRI (magnetic resonance imaging), which are preferred to ultrasound. The gold standard is venography, sometimes performed for surgical planning[2].

General features include absent infrarenal inferior vena cava with preservation of the suprarenal segment, common iliac veins may be absent, external and internal iliac veins drain into the ascending lumbar veins, which drain into the azygos-hemiazygos system or collateral paraspinal circulation may be present[3].

CASE REPORT

We present a case of a 20-year-old female patient with syncope who came to the emergency department. She had profuse perspiration, and she lost 12 kg of body weight in one month. Under the suspicion of pulmonary thromboembolism, a CT angiography of the pulmonary arteries showed a retrosternal para-aortic hypovascular soft tissue formation with a diameter of 30×40×85 mm. Laboratory findings also confirmed higher thyroid hormone values. Immunohyperthyroidism was diagnosed. A venous phase

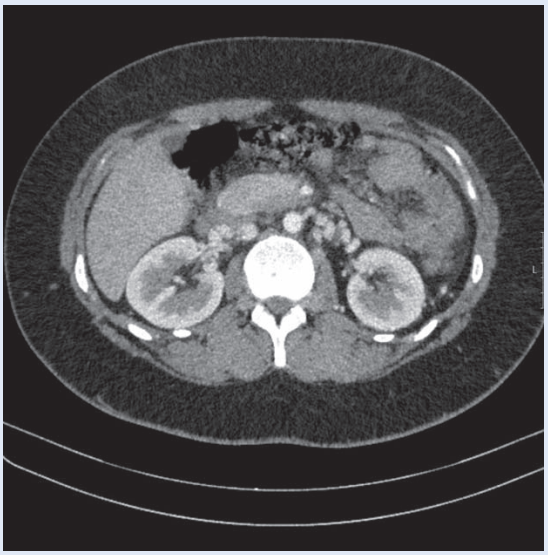

Figure 1. Axial abdominal CT scan – Absence of IVC.

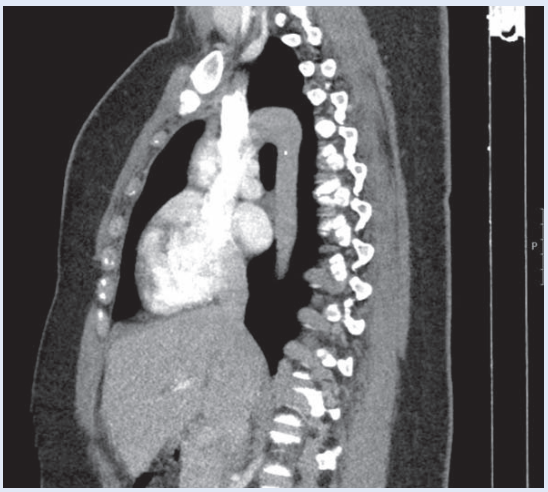

Figure 2. Sagittal abdominal CT scan – No representation of the inferior v. cava in the area from the renal veins to the heart.

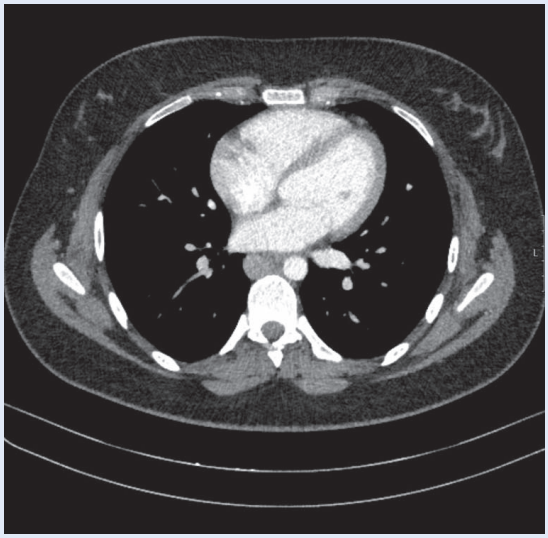

Figure 3. Axial abdominal CT scan – Hemiazygos vein merges into the azygos vein in the thorax area.

of CT angiograpy scan of the thorax was performed which showed that para-aortic hypovascular soft-active formation was the thymus, but an incidental finding was also discovered. There was no representation of the inferior v. cava in the area from the renal veins to the heart, and the dominant azygos and hemiazygos v. were visible. Hemiazygos vein merges into the azygos vein in the thorax area. All mentioned above corresponds primarily with the anatomical variant of congenital absence of v. cava inferior.

DISCUSSION

All mentioned above corresponds primarily with the anatomical variant of congenital absence of v. cava inferior. The development of the IVC is a very complex process involving multiple veins in the embryonic development. Understanding this developmental embryonic process is important for diagnosing and understanding congenital anomalies of the IVC.

The infrahepatic IVC develops from three paired embryonic veins at around 6-8 weeks of embryogenesis. Those three are posterior cardinal, subcardinal and supracardinal vein.

Absence of the infrarenal IVC would be the result of failure of the posterior cardinal and supracardinal veins to develop[1].

The collateral veins are enlarged with agenesis IVC, enabling blood transport to the superior vena cava. IVC agenesis (IVCA) may predispose to venous hypertension and complications, including thromboembolism[3].

There is an ongoing debate as to whether the absence of the IVC (either entire or infrarenal) is a true congenital anomaly or the result of perinatal IVC thrombosis. Thrombosis occurring around the time of birth could disrupt normal IVC development, leading to similar anatomical outcomes. Differentiating between congenital absence and acquired thrombosis is crucial for understanding the patient's condition and guiding appropriate management. Congenital absence often does not require aggressive treatment, except in cases of symptoms or recurrent thrombosis, whereas perinatal thrombosis is treated with anticoagulants, thrombolytic therapy, or even surgical intervention, depending on the severity and risk to the patient[4].

CT angiography is essential in detecting vascular malformations associated with IVCA. Incidental finding of IVCA in a patient initially suspected of having pulmonary thromboembolism. Lifelong anticoagulation therapy may be necessary in cases of IVCA with thrombotic complications.

Differentiating between these two conditions is crucial because a misdiagnosis could lead to overtreatment of an asymptomatic congenital condition or insufficiently aggressive management of acute thrombosis, which could endanger the patient's life.

CONCLUSION

IVCA is a significant but highly under-recognized cause of unprovoked DVT of the lower extremities in the young population without additional risk factors. Therefore, in unclear cases of unprovoked deep vein thrombosis of the lower extremities in younger population, this cause should also be considered, and accordingly, the necessary diagnostic evaluation should be performed, including imaging for vascular anomalies, alongside a full thrombophilia screening. The drainage of the lower limbs through the azygos vein might

be insufficient, causing stasis and eventually thrombosis. Treatment is mainly conservative and may require lifelong anticoagulation therapy.

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Conflicts of Interest: Authors declare no conflicts of interest.

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