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Case Report

Venous Thrombosis and Congenital Absence of Inferior Vena Cava in a Patient with Menorrhagia and Pelvic Pain

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Abstract. *Background:* Agenesis of the inferior vena cava (IVC) is an uncommon congenital vascular malformation. We report a case in a teenage female recently started on oral contraception.

Case: Because of menorrhagia, the patient had begun an oral contraceptive pill (OCP) 1½ months prior to presentation. She initially presented with pelvic and lower back pain, and imaging showed a pelvic deep venous thrombosis (DVT) and an interrupted IVC. Anticoagulation was started, the OCP was discontinued, and a discussion occurred regarding the treatment options for her menorrhagia following her recent diagnosis.

Summary and Conclusion: The case presented shows the rare occurrence of the congenital absence of an IVC with pelvic thrombosis in a young female with a history of menorrhagia and new onset of pelvic pain. The evaluation of this case report leads to a comprehensive review in the treatment choice for menorrhagia with the preceding history of a thrombotic event.

Key Words. Congenital—Interrupted—Absence—Inferior vena cava—IVC—Menorrhagia—Thrombosis—Deep vein thrombosis—DVT—Anticoagulation

Introduction

Agenesis of the inferior vena cava (IVC) is an uncommon congenital vascular malformation characterized by a developmental defect of the IVC and collateral circulation. The prevalence of congenital absence or interrupted IVC has been reported at 0.15%-3%.[1-3] The anomaly of an absent IVC is just 1 of multiple abnormalities that can occur during embryologic formation of the IVC, including transposition or duplication of the IVC, circumaortic venous rings, stenosis of the IVC with or without web formation, and retroaortic left renal vein.[1,4] The reported prevalence of all IVC anomalies ranges from 0.07%-8.7%.[5] These congenital abnormalities result from aberrant development of the IVC segments during the sixth gestational week of embryogenesis. During this time period, 3 paired veins (supracardinal, posterior cardinal, and subcardinal) normally fuse and regress, eventually forming the IVC.[5] Failure at this point of development leads to congenital abnormalities in the IVC, with the majority occurring above the level of the kidneys.[4] In patients with congenital absence of the IVC, alternative routes for venous return form; these alternative routes can include hemi-azygous, vertebrolumbar, anterior abdominal wall, and transumbilical portocaval collaterals.[4] Of interest, if these collateral vessel systems are well formed, patients tend to be asymptomatic.

Although in theory IVC absence may cause decreased venous return, leading to deep venous thrombosis, the association between the 2 is rarely reported. Ten cases of DVT in patients with absence of the IVC were reported between 1966 and 1999. [6] Tsuji reported on a 21-year-old male presenting with bilateral leg swelling, calf pain, and fever. Their review of the literature showed that all 10 previously reported cases occurred in young men with an average age of 29.8.[7] Yun presented a case in which a 39-year-old woman was diagnosed with congenital agenesis of the IVC in combination with hyperhomocysteinemia, leading to a DVT in her bilateral femoral veins.[8] The unique finding within this case was the rare occurrence of the absence of the infrarenal segment of the IVC, which is reported to occur in only 6% of IVC anomalies. The review performed by Yun showed 19 males and only 8 females diagnosed with

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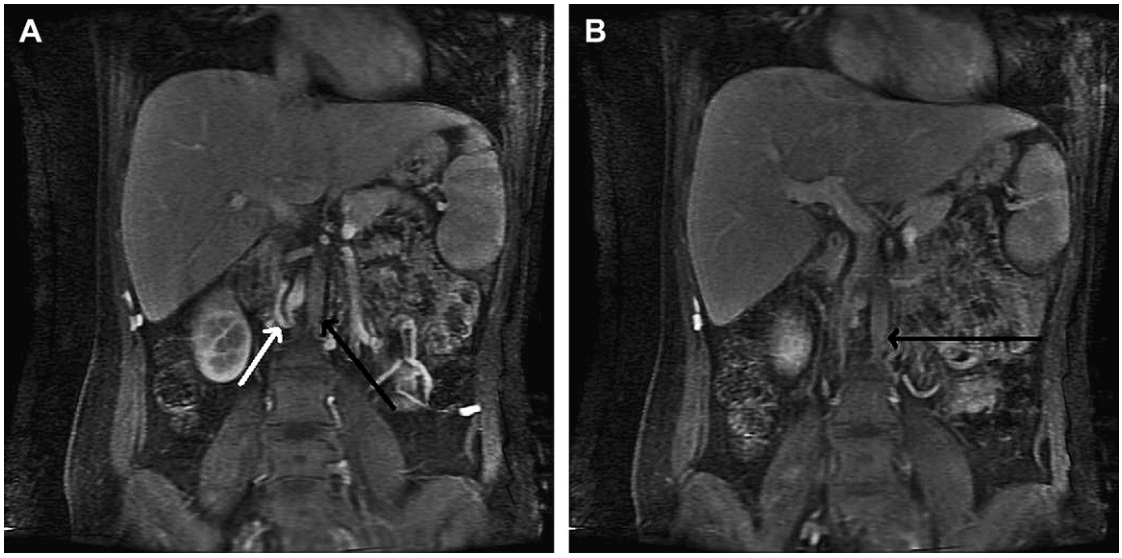

Fig. 1. MRI images demonstrating absence of inferior vena cava (IVC). (A) Image showing the superior aspect of IVC with the right collateral renal vein (White arrow) entering into the expected area of the IVC, with the stump appearance inferior to the entry point. Black arrow shows adjacent aorta. (B) Image demonstrates the aorta to the left of the IVC with the distal portion of the aorta continuing inferior and no visualization of an IVC. Black arrow showing descending aorta.

a DVT caused by congenital IVC malformations. Of the 27 reported cases, only 5 of the patients had an underlying coagulopathy and only 5 were noted to have precipitating factors, including immobilization, surgery, oral contraceptive pills (OCPs), trauma, and malignancy.[8]

Virchow described the 3 main causes leading to the formation of a thrombotic event: (1) abnormalities of the vessel wall; (2) blood components; and (3) dynamics of flow.[2] It has also been suggested that the infrarenal absence of the IVC could be attributed to intrauterine or perinatal thrombosis.[8] Overall etiologies of DVT are multifactorial and can include both congenital and acquired factors that lead to either hypercoagulability or venous stasis. An inadequate or abnormal blood flow through the collateral vessels may lead to an increase in the venous blood pressure in the lower extremities, altering dynamics of flow, and leading to a deep venous thrombosis.

Female reproductive-age patients on oral anticoagulation have reported significantly increased mean duration of menses and menorrhagia when compared to their menstrual cycles prior to the anticoagulation treatment.[9] Based on this information, one should consider the effect that anticoagulation therapy will have on a patient's menstrual cycle.

The case presented is important because it brings into account all of the following factors: the rare occurrence of congenital IVC anomalies, the risk of DVT in patients with absence of IVC, and the effect thrombosis and anticoagulation can have on female patients' menstrual cycles.

Case

An 18 year-old Caucasian female with an unremarkable medical history was transferred to our facility from a local hospital. She experienced menarche at age 12 with menses for 7 days, heavy enough to cause the patient to miss school. For this reason she had been started on a monophasic low-dose oral contraceptive pill (OCP) by her primary care physician 1½ months prior to presentation. The patient initially presented to a local emergency department with pain in her lower back and pelvis; the pain was worse with walking and was unrelieved by ibuprofen. A computed tomography (CT) scan at that time suggested a pelvic deep vein thrombosis (DVT) and an interrupted IVC. After being placed on low-molecular-weight heparin, she was transferred to our children's hospital for further evaluation and management.

There was no family history of thrombosis elicited. A hypercoagulability workup—including factor V Leiden, lupus anticoagulant, anticardiolipin antibodies, antithrombin III, protein C, protein S, MTHFR, homocystine, and prothrombin gene mutation—were all negative. Fibrinogen, D-dimer, and CRP were all elevated at 846 mg/dL, 1.07 μg/mL, and 150 mg/L, respectively. Magnetic resonance imaging and venography studies were performed and demonstrated agenesis of her inferior vena cava with multiple collateral vessels (Fig. 1). They also revealed a large clot in the left common and internal iliac veins with significant surrounding edema (Fig. 2). Multiple chronically enlarged veins were seen coursing