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Special Deep Vein Thrombosis in a Child: A Case Report

Xiaorong Wen[1] and Zhoupeng Wu[2,*]

[1]Department of Ultrasound, West China Hospital, 37 GuoXue Alley, Chengdu 610041, Sichuan Province, China
[2]Department of Vascular Surgery, West China Hospital, 37 GuoXue Alley, Chengdu 610041, Sichuan Province, China

*** *Corresponding author:*** Zhoupeng Wu, Department of Vascular Surgery, West China Hospital, 37 GuoXue Alley, Chengdu 610041, Sichuan Province, China. Tel: +8602885422870; Email: lzwzp19@163.com

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Abstract

Background: Agenesis of the inferior vena cava (IVC) is an extremely rare congenital malformation. Although frequently symptomatic, due to its low prevalence, agenesis of IVC may often be missed in the normal workup. Most of the reports are related to the loss of IVC alone, while the reports of simultaneous loss of IVC and bilateral iliac veins are rare. Acute deep venous thrombosis is also one of the causes of chronic deep venous ulcer, especially in children with acute deep venous thrombosis, which is an important cause of neglect and missed diagnosis. When color Doppler ultrasound cannot be clear, attention must be paid to computerized tomography examination.

Case Presentation: We reported a case of IVC below renal vein hypoplasia in an 8-year-old girl who presented bilaterally with persistent pain in both lower limbs. Ultrasonography revealed no clear IVC and iliofemoral venous system under the renal venous plane.

Conclusion: This case is a rare congenital malformation of IVC. It is rare in children with venous thrombosis-related symptoms. We intended to demonstrate the etiology of the appearance of venous thrombosis in children through this case.

Keywords: Agenesis, Inferior vena cava, Pediatric, Venous thrombosis

1. Background

Although venous thromboembolism (VTE) is a common clinical disease, it is mostly observed in adults. Recently, due to the gradual increase in the use of various catheters in children, the development of different imaging techniques, and the increase of clinical attention, the incidence of VTE in children has increased year-by-year (1). Congenital inferior vena cava (IVC) loss is a very rare vascular variant, and the first symptom of the disease may be deep vein thrombosis (DVT) of the lower extremities or accidentally found in ultrasound or computerized tomography (CT) screening. This is a very rare risk factor for DVT and is often ignored because it may be found without timely ultrasound or CT examination, especially for children. We reported a case of congenital absence of IVC and bilateral common iliac veins complicated with lower extremity DVT in a child.

2. Case Presentation

An 8-year-old girl was sent to the emergency department with continuous pain and swelling in her left lower limb for 1 week. The patient reported significant pain in the left groin area, accompanied by tenderness. The patient had no such symptoms before this admission. There was also no previous history of varicose veins and lower extremity ulcers. The patient had never underwent any surgery before, no taking special medications, and had a clear family history. Physical examination revealed significant swelling in the left lower limb (Figure 1A). Obvious varices were observed in her lower abdomen (Figure. 1B). Our clinical diagnosis was DVT of the left lower limb. Ultrasound showed DVT in the left femoral vein and external iliac vein. The patient's medical history did not suggest that she had a clear risk factor for DVT. Ultrasound showed (1) The absence of inferior hepatic IVC (Figure 2A), (2) The absence of the left common iliac vein, the junction of the left external iliac vein (thick arrow), and internal iliac vein (thin arrow) (Figure 2B), (3) The absence of the right common iliac vein, the blood flow of the right external iliac vein flows to the internal iliac vein (as shown by the arrow in the figure), and the blood flow of the internal iliac vein is reversed (Figure 2C). (4) A tortuous compensatory vein was found on the

Figure 1. Significant swelling of the left lower limb are revealed through physical examination (Figure 1A). Obvious varices in the lower abdomen (Figure 1B).

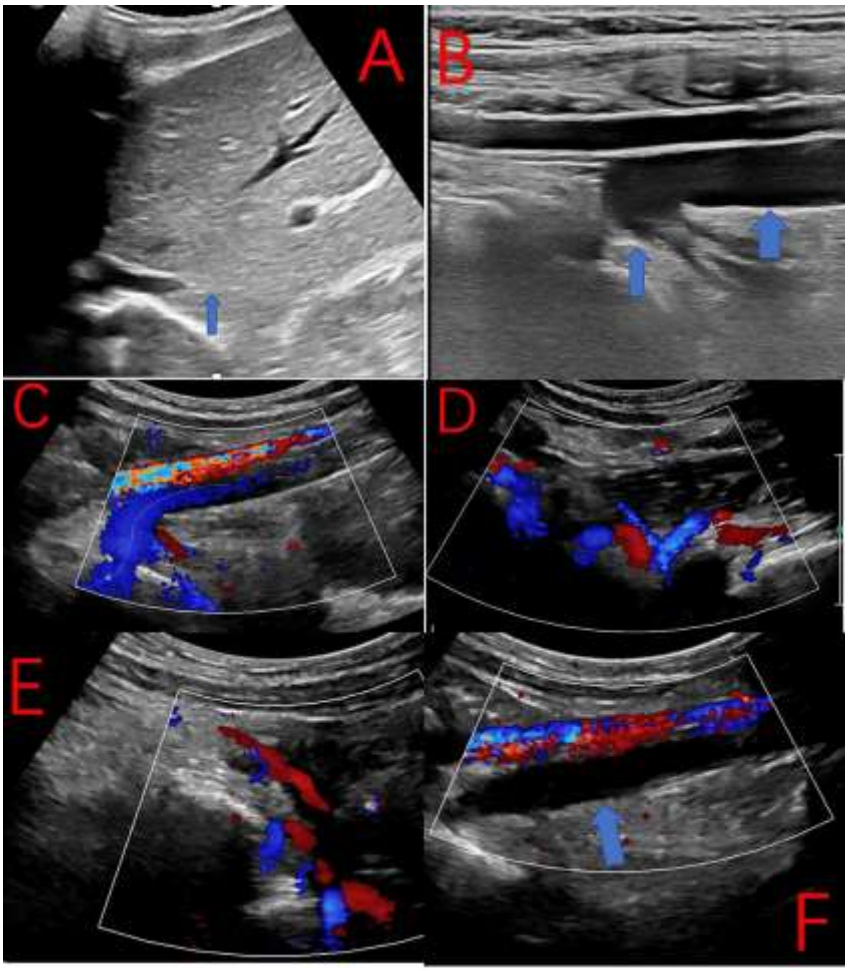

Figure 2. Absence of inferior hepatic inferior vena cava (Figure. 2A), Absence of left common iliac vein, the junction of left external iliac vein (thick arrow) and internal iliac vein (thin arrow) (Figure. 2B), Absence of right common iliac vein, blood flow of right external iliac vein flows to internal iliac vein (as shown by the arrow in the figure), and blood flow of internal iliac vein is reversed. (Figure. 2C), A tortuous compensatory vein is found on the deep surface of the psoas major muscle near the right spine (Figure. 2D), A distorted compensatory vein is found near the left spine (Figure. 2E), A weak echo filling is found in the lumen of the left external iliac vein, and no blood flow signal is found (Figure. 2F).

deep surface of the psoas major muscle near the right spine (Figure 2D), (5) A distorted compensatory vein was found near the left spine (Figure 2E) (6) A weak echo filling was found in the lumen of the left external iliac vein, and no blood flow signal was found (Figure 2F). (7) There was no obvious abnormality in the abdominal aorta. There was no evidence of thrombosis in IVC and bilateral common iliac veins. According to the mentioned physical and ultrasonic examinations, it was diagnosed that the patient suffers from left lower extremity DVT due to the congenital absence of IVC and bilateral common iliac veins. After five days of treatment with low molecular weight heparin, the symptoms were completely relieved. Then, the patient received anticoagulant therapy with warfarin for one year and long-term compression therapy. The child was in good condition during the one-year follow-up and had no symptoms of recurrent DVT or lower extremity venous ulcer.

3. Discussion

Deep vein thrombosis is common in adults and rare in children, often due to the increased central venous access equipment and the significant progress of child management under chronic and/or severe diseases. Congenital absence of IVC is considered to be a very rare risk factor for DVT in children. Published studies were searched in MEDLINE from 1960 to 2009 and only 23 cases were found (3-24); this number may be underestimated because conventional ultrasound might be unable to detect the abnormalities of IVC and iliac vein. There is also a literature report that the incidence rate of vascular dysplasia is up to 5% in hospitalized patients aged < 40 years (1,2), and they are often diagnosed as idiopathic DVT (3-5).

The infrarenal IVC develops from three paired veins at 4-8 weeks of gestation: the posterior cardinal, subcardinal, and supracardinal veins. These

form the IVC below the retrohepatic portion, along with the azygous and hemiazygous systems and the iliac veins. The absence of the infrarenal portion of the IVC is among the rarest of the anomalies described and is caused by failure of the posterior cardinal and supracardinal veins with preservation of the suprarenal segment. The part of IVC above the kidney vein is formed by the subcardinal vein. Due to the formation of extensive collateral circulation network, the absence of deep venous system can be compensated, so the symptoms caused by vascular hypoplasia may be asymptomatic or occult. Despite the existence of these collateral circulations, the venous reflux of lower limbs is still insufficient, which may still lead to venous blood stasis and DVT (6).

By searching previously published cases, we found some common clinical manifestations of these patients. The clinical manifestation of most cases is proximal DVT, often involving iliofemoral vein. Some patients will have secondary venous ulcer. The patients with DVT associated with this type of vascular hypoplasia are younger, even children. Compared with the current international reports, the age is < 30 years (7, 8). The vast majority of cases showed DVT in both lower extremities, and less single lower limb DVT. At the same time, so far, no such cases of vascular hypoplasia are associated with potential malignant tumors in vivo. Like the chlid in this study, in the vast majority of these patients, abdominal CT or magnetic resonance imaging (MRI) can often detect the formation of a large number of collateral branches in the lumbar spine and beside the spine, and the azygos and semi azygos veins that expand the drainage of iliac veins. There are a few reports that these enlarged azygos and semi- azygos veins will become abnormal channels causing a pulmonary embolism (9-12).

Around one-third of cases were diagnosed with deep and superficial venous thrombosis (13, 14). Some scholars believe that this congenital absence of IVC is secondary to perinatal thrombosis, resulting in the regression of normal IVC (15). There is no evidence to support this theory in the history or imaging examination of this child. The formation of collateral circulation network is insufficient, which cannot provide enough channels for blood reflux, resulting in increased venous pressure and stasis. It has been recognized as an important risk factor for the DVT development in such patients and has led to clinical symptoms in six patients. Other congenital abnormalities involving hypoplasia of the right kidney were diagnosed in six patients. The occurrence of this condition can be explained by the corresponding theory of embryology, since the reflux of the right renal vein relies mainly on the IVC. Therefore, complete or partial absence of IVC may affect renal development, especially in children (13,14).

Color Doppler flow imaging is the first choice for all patients with suspected DVT. It can show the iliac vein and IVC. However, it may miss some IVC cases due to asymptomatic or ultrasound technology. The CT or MRI can easily diagnose such patients. According to the current literature, it is suggested that for young patients, if the ultrasound does not fully display the IVC or iliac vein, and there are no clear risk factors for iliac-femoral vein thrombosis in the lower extremities, abdominal CT should be performed to exclude this kind of vascular dysplasia (12,14). At the same time, patients should also be screened for thrombotic diseases (e.g., thrombophilia). Although all patients in the existing literature took oral anticoagulants, there was no evidence of the best treatment time. However, two cases of DVT recurrence were reported after one and two years of treatment. Due to the particularity of the case, there are currently no related studies on the long-term follow-up of such patients. Such patients should also be followed up clinically to prevent late Post-thrombotic syndrome (PTS) (3,5,12,25-28).

So far, surgical revascularization has been used to treat intractable ulcers that fail to respond to conservative treatment. The effectiveness of this procedure was confirmed by artificial reconstruction of the IVC and an artificial graft bypass from the iliac vein to the internal thoracic azygos vein[7-8]. All young patients with DVT, especially children, should consider the presence of this type of hypoplasia; consequently, an ultrasound of the IVC and iliac vein is necessary to exclude the hypoplasia while completing skin and subcutaneous blood vessel-related examinations. If ultrasound cannot find the IVC or iliac vein, these patients need abdominal CT to find the possible cause of DVT.

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Footnotes

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