

An unusual anomaly of deep venous system in the lower limb: Complete unilateral agenesis of iliofemoral veins in the absence of persistent sciatic vein

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ABSTRACT

Venous anomalies are the most common congenital vascular anomalies. The rarest of these is complete deep venous agenesis. Here, we report a case of a 15-year-old male patient evaluated on color Doppler and computed tomographic venogram with complete right iliofemoral deep venous agenesis with the inferior vena cava continuing as the left common iliac vein and the right popliteal vein draining into superficial anomalous veins of the thigh. These superficial veins were seen to drain through a superficial suprapubic arcuate connector vein into the dilated left common femoral vein. Uniquely in our case, there was no persistent embryonic sciatic vein and no demonstrable reflux into the superficial connector vein, suggesting an uncommon embryological mechanism underlying its pathogenesis. Surgical resection of the dilated superficial veins is absolutely contraindicated in cases of deep venous agenesis and hence accurate diagnosis by the radiologist is absolutely essential in such cases.

Key words: *Congenital venous anomaly, Deep venous agenesis, Ilio-femoral venous agenesis, Persistent sciatic vein.*

The deep venous system of the lower limbs has relatively constant anatomy. However, among congenital vascular anomalies, venous anomalies predominate and deep veins of the lower limb are common sites of such anomalies. Common variations include phlebectasia, venous aneurysms, duplications and variations in the course of the veins while a relatively uncommon variant includes a valvulosis. The rarest of the congenital anomalies of deep veins is agenesis or complete absence of veins [1]. Congenital deep venous anomalies have also been reported to be more common in the left as compared to the right lower limb [2]. Such cases usually present early in childhood with extensive varicose veins, deep venous thrombosis or complications secondary to chronic venous insufficiency in the lower limb.

Here, we report a rare case of an unusual anomaly of the right lower limb deep venous system with complete agenesis of the right iliofemoral veins and continuation of the inferior vena cava (IVC) as the left common iliac vein. While this anomaly in itself has only been reported a few times in literature, reported cases of complete iliofemoral venous agenesis almost always have a persistent embryonic sciatic vein providing venous drainage for the lower limb [2,3,4]. However, in our case the embryonic sciatic vein was also absent; anomalous dilated superficial veins provided the only venous drainage for the right lower limb.

CASE REPORT

A 15-year-old male patient presented to the department with complaints of prominent superficial veins in the right lower limb

which had progressed over the past eight years, with associated heaviness which became more prominent with activity. He had no complaints of any obvious swelling, ulcerations or other skin changes in the limb. He was a school student and was not involved in any activity that necessitates prolonged standing. No similar complaints were present in any of his family members.

On examination, the patient's general condition was fair and vitals were stable. The right lower limb showed prominent dilated, non-tortuous superficial veins involving the thigh and the right lower abdominal wall in the regions of the right iliac fossa and hypogastrium extending to the left iliac fossa. They were seen to end abruptly just distal to the left inguinal ligament (Fig. 1). Right lower limb length and girth were normal. The left lower limb was normal. In view of the unusual appearance and distribution of dilated superficial veins, possibility of an anomalous deep venous system involving the right iliofemoral veins was suspected and Doppler evaluation of the right lower limb was performed.

On Doppler evaluation, the right common iliac vein was not visualized from the expected point of its origin at the distal end of the IVC. The right external iliac, common femoral, superficial femoral and deep femoral veins were also not visualized. The right popliteal vein was visualized, as were the deep veins of the right leg distal to the popliteal vein (Fig. 2). The proximal end of the popliteal vein was found to communicate with superficial veins in the thigh. The direction of flow was from deep veins to superficial veins. The embryonic sciatic vein, which was expected to be persistent, was also not visualized. The great saphenous vein, as well as other anomalous superficial veins, were seen to



Figure 1: The patient showing dilated superficial veins in the right thigh (A), suprapubic region (B) and abruptly ending just below the level of the left inguinal ligament (C).

be dilated, non-tortuous, and were found to drain into a large superficial suprapubic arcuate connector vein in the subcutaneous plane of the anterior abdominal wall. The connector vein was found to cross the midline and drain into the left common femoral vein which was dilated. Only forward flow from the connector vein into the left common femoral vein was observed without any reflux (Fig. 3). The IVC was found to continue into a dilated left common iliac vein which was then seen to give rise to a dilated left external iliac vein and left common femoral vein with which the superficial connector vein from the right lower limb was seen communicating. The arteries of the right lower limb were found to be normal in course and anatomy, albeit without the accompanying major veins up to the level of the distal superficial femoral artery.

Doppler evaluation was followed by a timed computed tomographic venogram (CTV) with intravenous contrast administration for complete evaluation of veins in deeper intermuscular planes not accessible to Doppler evaluation, as well as to identify any additional anomalies not detected on Doppler examination. Just distal to the aortic bifurcation, the IVC at its distal end was seen to cross from its normal right paramedian

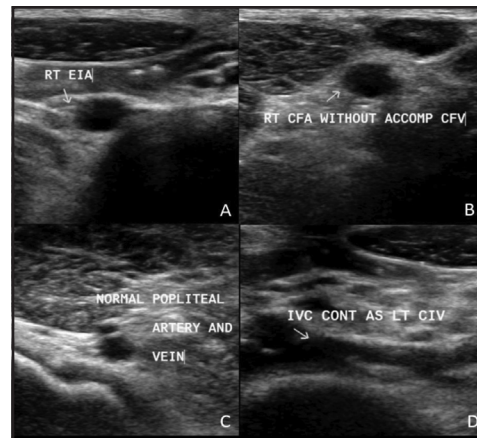


Figure 2: Doppler evaluation of the patient. The right external iliac vein (A) and the right common femoral vein (B) are not visualized alongside their respective arteries. The right popliteal vein is visualized in the popliteal fossa (C). The IVC is seen to continue directly as the left common iliac vein (D).

position to the left side posterior to the bilateral common iliac arteries and continue along the left common iliac artery following the course of the common iliac vein and its further divisions. Left internal and external iliac veins, as well as the left common femoral vein to the drainage of the suprapubic connector vein, were dilated (Fig. 4).

The dilated superficial veins and suprapubic arcuate connector vein were well visualized on multiplanar maximum intensity projection (MIP) images (Fig. 5). No evidence of any linear structure resembling normal or abnormal deep veins was found accompanying the arteries of the right lower limb from the level of the common femoral artery up to the level of the distal superficial femoral artery, suggesting complete agenesis of these veins. The whole of the IVC up to the right atrium was normal. Arterial system of bilateral lower limbs was also normal. Rest of the findings seen on Doppler examination were confirmed. No other additional vascular malformations or anomalies were observed.

In view of complete visualization of the entire vascular system on CTV and high risk of venous gangrene in the event of thrombosis of the connector vein, invasive venography was

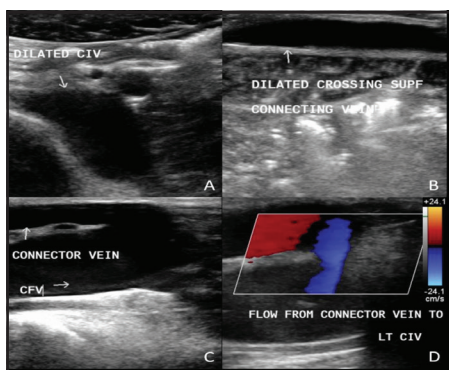


Figure 3: Doppler evaluation of the patient. The left common iliac vein is dilated (A). A dilated suprapubic connector vein is seen crossing the midline from right to left in the subcutaneous plane (B) and draining into the dilated left common femoral vein (C) with unidirectional flow from the connector vein to the left common femoral vein (D).

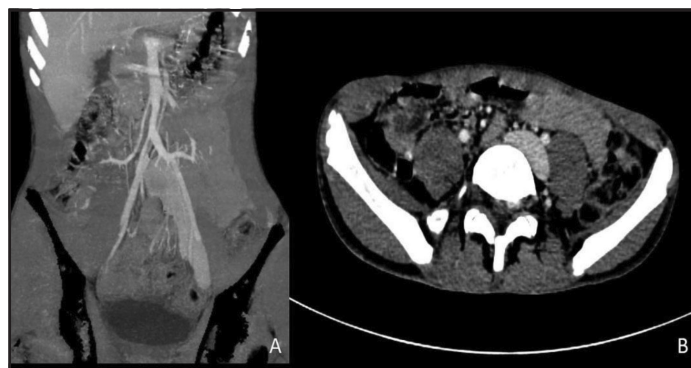


Figure 4: CTV of the same patient. Coronal MIP image of the major abdominal vessels (A) and an axial section of the lower abdomen at the level of the iliac bones (B) show a direct continuation of the IVC as the dilated left common iliac vein and subsequently left external iliac vein accompanying the respective arteries.



Figure 5: CTV of the same patient. Coronal MIP images of the right thigh and lower suprapubic abdomen show dilated non-tortuous superficial veins (A), which drain into a superficial arcuate connector vein passing in the suprapubic region of the anterior abdominal wall from right to left (B) to drain into the dilated left common femoral vein accompanying the left common femoral artery (C). The absence of any deep vein accompanying the right superficial femoral artery can also be appreciated (C).

not performed. As the patient did not have any significant complications of chronic venous insufficiency, surgical placement of venous grafts was deemed unnecessary at this stage. The patient was conservatively managed, prescribed compression stockings and is currently on follow-up for any progression of symptoms.

DISCUSSION

Prevalence of various congenital vascular malformations was studied in detail by Eifert S *et al.* The overall prevalence of congenital vascular malformations is about 1.5%. Among these, venous malformations are most common (two-thirds), of which deep venous malformations comprise 47%. Deep venous malformations include phlebectasia, venous aneurysms, duplications, variant course, avalvulia, and venous hypoplasia or aplasia. Venous aplasia was found to be the least common among these (2%) in their study [1].

Venous anomalies of the lower limb can usually be suspected through a thorough clinical examination of the patient. However, a complete characterization of the anomalies invariably necessitates imaging. Doppler evaluation can provide information on the location and extent of the anomalies, but a complete evaluation of the anomalous venous system, especially in cases of agenesis of major veins, requires angiographic techniques in order to image the venous system. In the majority of the cases, complete evaluation can be performed using computed tomographic venography (CTV) or magnetic resonance venography (MRV) with appropriate reconstructions without the need for invasive angiography [2].

For a complete understanding of the congenital venous malformations of the lower limb, knowledge of embryogenesis of the lower limb venous system is necessary. The lower limbs develop between the 4th and 8th postovulatory weeks. Embryological vessels follow a pattern of rapid development and involution as they are progressively replaced by the definitive venous system. Any aberration in this process can lead to the development of congenital venous malformations. The specific malformation depends upon the stage at which the defective development occurs.

Lee BB has described the development of the lower limb deep venous system in three distinct phases. In the first phase, drainage of the lower limb occurs through the primitive fibular vein (the first embryonic vein) into the posterior cardinal vein. In the second phase, the sciatic vein (the second embryonic vein) is formed and consists of the anterior tibial and the primitive fibular veins together, passing along the dorsal aspect of the limb to drain into the posterior cardinal vein. In the third phase, the primitive femoral vein (the third embryonic vein) is formed from the posterior cardinal vein and grows distally. The primitive femoral vein and a connecting vein together form the definitive femoral vein. The distal part of the sciatic vein persists to form the definitive posterior tibial veins while the proximal part of the sciatic vein in the thigh regresses. The posterior cardinal veins eventually form the definitive iliac veins [3].

A defect in the distal part of the posterior cardinal vein leads to agenesis of the iliac veins and proximal part of the femoral veins. A defect in the transition from second to the third stage results in persistent marginal vein while a defect in the transition beyond third stage results in persistent sciatic vein with defective formation of the femoral vein. Agenesis of the femoral vein is hence closely associated with persistent sciatic vein [3, 4]. Our case is unique in this aspect that in spite of complete agenesis of the right iliac and femoral veins the sciatic vein was not found to be patent.

Based on this, we have hypothesized that the pathogenesis in our case could be some intrauterine insult to the deep venous system of the lower limb after the third phase of venous development (after involution of the embryonic sciatic vein), which in turn could have led to early involution of the definitive major deep veins with compensatory enlargement of superficial veins and development of anomalous superficial veins for venous drainage of the right lower limb. Another factor in support of our hypothesis is the fact that in most cases of congenital venous aplasia with persistent embryonic veins, unilateral limb hypertrophy with edema and tortuous superficial varicosities are a very common feature. This is because persistent embryonic veins lack venous valves which leads to severe reflux and chronic venous insufficiency [5]. The absence of demonstrable reflux into the superficial suprapubic arcuate connector vein in our case implies the lack of persistence of embryonic veins which again suggests pathological involution of the definitive deep veins after physiological involution of embryonic veins in intrauterine life.

The major syndromic association with deep venous agenesis is Klippel-Trenaunay-Weber syndrome. This syndrome is highly variable in presentation, with a wide variety of features being considered under its spectrum [6]. Oduber CE *et al.* in 2008 reviewed the existing literature on the syndrome and proposed diagnostic criteria for the same [7]. Our patient did not fit into the diagnostic criteria for this syndrome.

The primary pathophysiological mechanism underlying clinical presentation of deep venous agenesis is chronic venous insufficiency with venous stasis in the limb due to

poor outflow through superficial veins and reflux through the valveless persistent embryonic veins. Subjective symptoms of patients include pain, heaviness, and swelling of the involved limb. Objective clinical signs include telangiectasias, tortuous and dilated varicosities, edema, pigmentation and eczema, lipodermatosclerosis and venous ulcers [8]. A key feature that points to the need for careful evaluation of the deep venous system in these cases is the presence of suprapubic dilated superficial veins, which occur only in patients with non-patent iliac veins, either secondary to deep vein thrombosis or, as in our case, agenesis of the iliac vein.

Although cases of lower limb deep venous agenesis are very rare, accurate diagnosis is vital in planning management. Failure to evaluate the lack of deep venous system patency in such cases may lead to misdiagnosis of the condition as primary venous insufficiency with superficial varicose veins due to incompetent perforators. This would result in inappropriate surgical planning, and extensive resection of the dilated superficial veins, leading to severe venous insufficiency which may culminate in venous gangrene of the limb. Surgical resection of the superficial veins is, therefore, contraindicated [9]. The recommended line of management includes the prescription of compression stockings started as early as possible with appropriate lifestyle changes [10].

CONCLUSION

While deep venous agenesis is in itself a very rare congenital anomaly, we have reported a unique case where both the definitive deep veins as well as the embryonic veins of the right lower limb were absent from the level of the common iliac vein to the level of the distal superficial femoral vein. Accurate diagnosis in such unusual cases is vital as surgical resection of the dilated superficial veins is absolutely contraindicated and can result in

disastrous consequences if accidentally performed following a failure to diagnose the absent deep veins.

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