

# ABDOMINAL WALL VARICOSITIES SECONDARY TO ONE-SIDED ILIAC VEIN AGENESIS

## TEK TARAFLI İLİAK VEN AGENEZİSİNE BAĞLI KARIN DUVARI VARİSLERİ

Özlem Barutçu SAYGILI, M.D.,  
Mehmet ÇOŞKUN\*, M.D.

N. Çağla TARHAN\*, M.D.,

Banu ÇAKIR\*, M.D.,

Baskent University Faculty of Medicine, Department of Radiology, Adana Teaching and Medical Research Center, Adana, Turkey  
Baskent University Faculty of Medicine, Department of Radiology\*, Ankara, Turkey  
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**SUMMARY:** Spiral computed tomography incidentally revealed one-sided iliac vein agenesis in two patients. In the first patient CT showed left-sided common iliac vein and external iliac vein agenesis, and in the second patient left-sided common iliac vein agenesis and left external iliac vein hypoplasia. In addition to these findings, both patients had abdominal wall and suprapubic varicosities without agenesis of the inferior vena cava. This has been documented in only one patient previously in the literature. Spiral CT is an accurate way to identify iliac vein agenesis and associated varicosities.

**Key Words:** Abdominal Wall Varices, Agenesis of Iliac Veins, Spiral CT.

### INTRODUCTION

Iliac vein agenesis is a very rare congenital anomaly (1,2) that is most often detected as a component of Klippel-Trenaunay syndrome (1-3). When iliac vein agenesis occurs, it is usually seen in conjunction with agenesis of the inferior vena cava (4-7). The clinical presentation varies according to the patency of the collateral veins (3). Iliac vein agenesis is usually identified in infancy or childhood (7). Most cases have been definitively diagnosed by ascending phlebography (2-4). In this report, we describe the spiral computed tomography (CT) features of two cases in which the anomaly was detected incidentally. Both patients showed one-sided iliac vein agenesis, and lower anterior abdominal and suprapubic varicosities.

**ÖZET:** İki hastada BT de rastlantısal olarak tek taraflı ana iliak ven agenezisi saptandı. Hastaların ilkinde spiral BT de sol ana iliak ve eksternal iliak ven agenezisi, diğerinde sol ana iliak ven agenezisi ve sol eksternal iliak ven hipoplazisi gösterildi. Bu bulgulara ek olarak her iki hastada inferior vena cava agenezisi olmaksızın karın ön duvarı ve suprapubik varisler bulunmaktaydı. Belirtilen bulgular daha önce sadece bir hastada tanımlanmıştır. Spiral BT iliak ven agenezisini ve eşlik eden varisleri saptamada güvenilir bir yöntemdir.

**Anahtar Kelimeler:** Karın Duvarı Varisleri, İliak Ven Agenezisi, Spiral BT.

### CASE REPORTS

#### Case 1.

A 68-year-old man was referred to our CT department for regular follow-up of rectal carcinoma. Scanning was performed with single-slice spiral CT (Somatom Plus 4, Siemens, Erlangen) in 5-mm-thick sections before and after bolus administration of intravenous contrast. The CT images revealed dilated, tortuous, venous collaterals in the lower anterior abdominal wall and suprapubic region (Fig. 1a). The inferior vena cava (IVC) appeared normal throughout its course (Fig. 1b), but the left common iliac vein was not detected (Fig. 1c). A small vascular structure posterior to the left external iliac artery was suspected to be a hypoplastic left external iliac vein (Fig. 1a). We

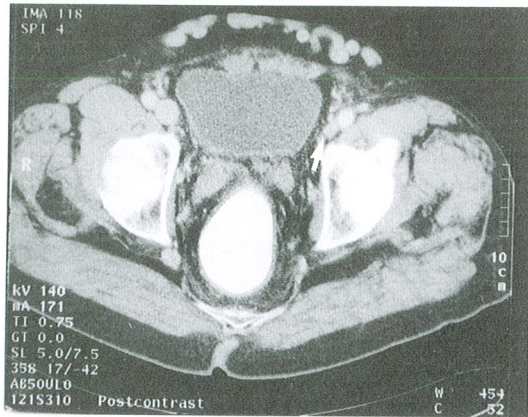


Fig. 1A: Axial post-contrast CT images of the first case: Dilated, tortuous, venous collaterals were demonstrated in the subcutaneous tissue of the lower anterior abdominal wall and in the suprapubic region. The small vascular structure posterior to the external iliac artery (short white arrow) was suspected to be the hypoplastic left external iliac vein.

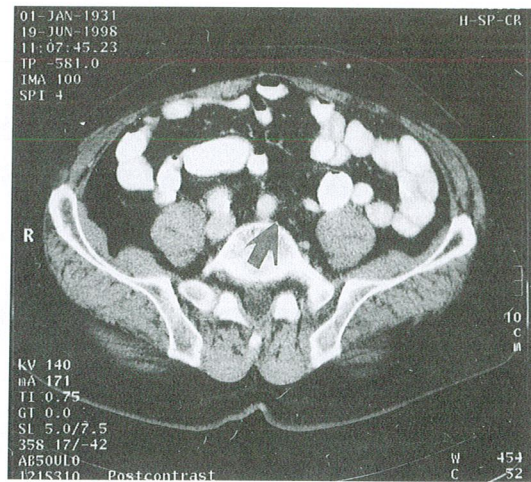


Fig. 1C: A CT image of the lower abdomen shows no common iliac vein on the left side (black arrow).

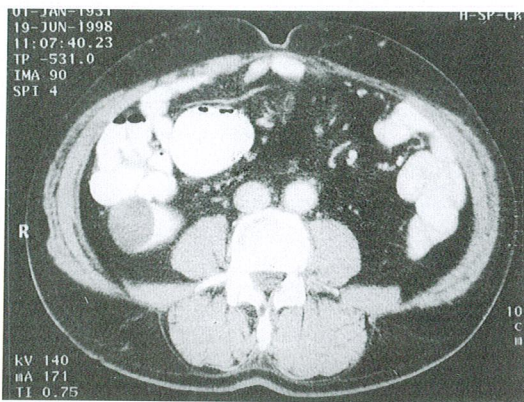


Fig. 1B: Inferior vena cava was normal.

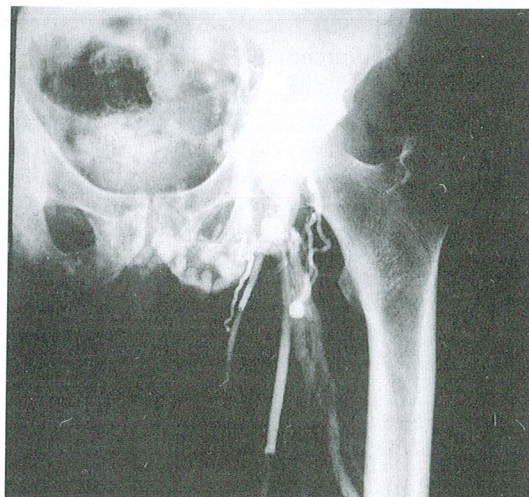


Fig. 1D: On lower extremity phlebography left common and external iliac vein were not detected, dilated tortuous venous collateral veins were seen in the anterior abdominal region.

also observed a vascular channel connecting the left femoral vein and the abdominal collaterals at the level of the symphysis pubis on the left side. The right common and external iliac veins were normal (Fig. 1a, 1c). The patient stated that such vascular dilatations had been present since childhood. The vessels had gradually dilated and become more tortuous over the years. The patient had no hemangiomas or any limb hypertrophy, thus ruling out a diagnosis of Klippel-Trenaunay syndrome. Based on these findings, he was

diagnosed with left common iliac vein agenesis accompanied by collateral varicosities. The findings were confirmed with venography (Fig. 1d, 1e).

#### Case 2.

A 76-year-old man with hypertension was referred to our radiology department for investigation of an abdominal aortic aneurysm. Left-sided lower limb hypertrophy was noted on physical examination. Spiral CT scanning was performed in 8-mm-thick sections after intravenous contrast injection. The images

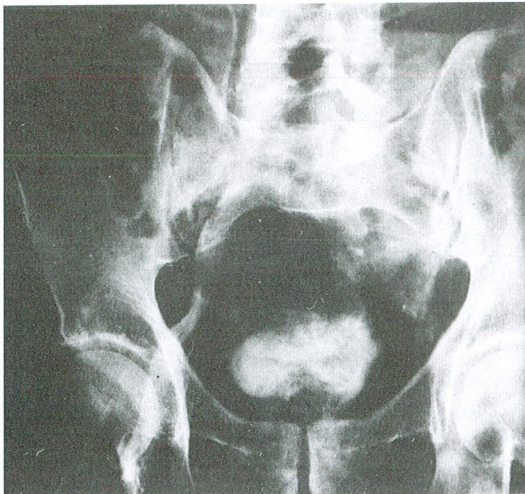


Fig. 1E: The right sided iliac veins were normal.

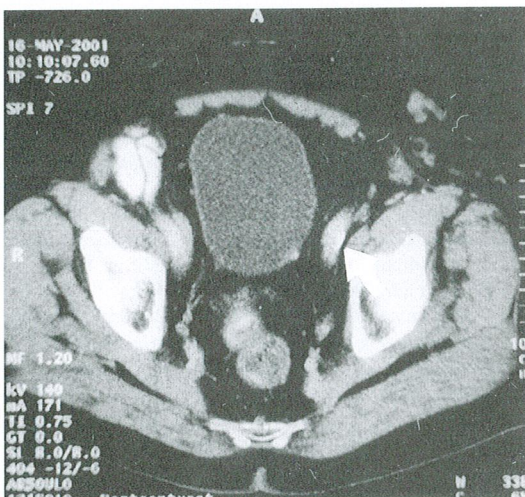


Fig. 2: Axial postcontrast CT image of the second case showing anterior abdominal wall collaterals, the left iliac vein was not present.

showed dilated, tortuous, venous collaterals in the lower anterior abdominal wall and suprapubic region (Fig. 2). The left common and external iliac veins were absent and the IVC appeared normal. The patient had had varicosities in his left thigh and leg since childhood, and this limb had been hypertrophic since that time. He had developed anterior abdominal and suprapubic varicosities in early adulthood. There were no cutaneous hemangiomas on physical examination and so Klippel-Trenaunay syndrome was ruled out.

## DISCUSSION

Although common congenital variations of the iliac veins and IVC have been described previously (8), iliac vein agenesis is a very rare congenital abnormality (1,2). Absence of the infrarenal segment of the IVC and iliac veins without any symptoms has recently been reported in the literature (4,7). To our knowledge, one-sided iliac vein agenesis without agenesis of the IVC or any associated congenital anomalies has been documented in only one patient prior to this report (1).

Two different pathogenetic mechanisms have been proposed to explain this form of vascular agenesis. The first is the occurrence of a mesodermal abnormality at 3-6 weeks of gestation. The second is the development of iliac vein thrombosis and IVC thrombosis during the intrauterine period (3,4). The embryologic development process of the iliac veins is complex and still not completely understood (1,2). The first venous system that develops in the embryo is that of the cardinal veins. In this arrangement, the anterior cardinal veins drain the cranial part of the embryo, and the posterior cardinal veins drain the caudal section. When these vessels become obliterated, the subcardinal veins appear, followed by the supracardinal veins. The supracardinal veins are connected to each other, and also connect with the subcardinal veins. Eventually, the right supracardinal vein becomes the infrarenal segment of the IVC, and anastomosis of the right subcardinal and supracardinal veins forms the renal segment of the IVC (1,2,9). A connection between the right and left supracardinal veins forms the left iliac vein. In this configuration, blood drains from left to right. If the anastomosis process fails, the result is agenesis of the left iliac vein (1,2). In the first patient, suprapubic varicosities were noted in childhood. The other patient showed leg varicosities in childhood. The CT scans showed normal IVC in both cases. These features are in accordance with the second etiological theory above.

The varicose veins that develop secondary to deep vein agenesis are important. If a patient shows no symptoms of venous insufficiency, then the varicosities are adequately draining the extremity (1,2,7). Typically, these vessels develop along the lateral aspect of the thigh in the

involved extremity, as was seen in our second case (2). Lower abdominal and suprapubic varicosities are also common (1,2,7). It is extremely important that the etiology of such varicosities be investigated before any decision is made to remove them for cosmetic reasons (1,7).

Numerous reports have discussed the definitive identification of this entity by phlebography (1-4). Spiral CT allowed non-invasive identification of the congenital anomaly in both of our patients, and these are the first two cases of this kind in which spiral CT has demonstrated the abnormality clearly (4,5,7).

Although phlebography was performed for confirmation in the first patient, the CT images clearly showed that there was no common iliac vein on the left side in this case. CT also provided detailed information about the location of the patient's varicosities, and revealed a small venous structure that was suspected to be the hypoplastic external iliac vein. This venous structure was thought to be the hypoplastic external iliac vein because CT demonstrated collaterals draining from the femoral vein. In addition, small collaterals that were not visible on venography were easily visualized on the spiral CT images. In the second case, the diagnosis was made without phlebography confirmation.

A further unique feature of both these cases is that iliac vein agenesis was diagnosed incidentally in late adulthood. Such abnormalities are usually identified before adolescence, when the individual develops symptoms of venous circulatory insufficiency.

For differential diagnosis, acquired left iliac vein stenosis or occlusion due to compression (Cockett syndrome) should be kept in mind in these patients (10). However, no venous structure was recognized in place of iliac veins in our patients, and there was no anatomical or pathological structure that would cause compression of these vessels to become stenotic or occluded.

In conclusion, congenital one-sided iliac vein agenesis not associated with other congenital anomalies is extremely rare. To our knowledge, as mentioned before, this is the second report of isolated one-sided iliac vein agenesis without other congenital anomalies. Spiral CT is a valuable non-invasive method that can be used to

diagnose this anomaly and associated varicous venous collaterals.

**Correspondence to:** Özlem BARUTÇU SAYGILI  
Çınar Sitesi 1.blok No:19  
Ümitköy  
06530 ANKARA - TÜRKİYE  
Phone: 312 236 10 55  
Fax: 312 221 32 76  
E-mail: obarutcu@yahoo.com

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