

# Thrombosed iliac venous aneurysm: A rare form of presentation of a congenital anomaly of the inferior vena cava

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Aneurysms of the iliac veins are extremely rare. We report a case of a 51-year-old male who was admitted for swelling of the lower right limb. Sonography and ascending phlebogram showed a complete occlusion of the right femoropopliteal veins, both iliacs and the inferior vena cava (IVC). A large collateral circulation throughout the paravertebral plexus and azygos system was also observed. The CT scan revealed a 5 × 9cm thrombosed aneurysm of the right external iliac vein and a congenital hypoplasia of the infrarenal IVC. Anticoagulant treatment and compression with elastic stocking was started. The 3-month follow-up showed mild residual edema of the right lower limb. The literature on this pathology is extensively reviewed. (*J Vasc Surg* 2008;48:218-22.)

The iliac vein is the least frequently reported location for aneurysms of the venous system. We present an unusual case of an iliac aneurysm secondary to a congenital anomaly in the drainage of the inferior vena cava (IVC). The literature on this pathology is extensively reviewed and analyzed.

## CASE REPORT

A 51-year-old male patient presented with fever, pain, and edema of the lower right limb for the last 3 days. The patient did not have any past medical or surgical history of interest. Additionally, history of varicose veins or trauma and presence of respiratory symptoms suggestive of pulmonary thromboembolism (PTE) were denied. On physical examination, a global edema of the right lower limb with severe cyanosis was notable. Distal pulses were present. The rest of the examination was normal except for moderate varicocele.

A venous Doppler ultrasound of the lower limbs followed by an ascending phlebogram were performed, showing a complete occlusion of the right femoropopliteal veins, right external iliac, both common iliacs, and the IVC. A large collateral circulation throughout the paravertebral plexus and azygos system was observed (Fig 1).

An abdominal 64-slice angio-CT scan showed thrombosis of the IVC and both common iliac veins. A 5 × 9 cm aneurysm of the right external iliac vein and a 2.5 cm ectasia on the left iliac were observed. Additional finding included a hypoplasia of the infrarenal IVC at its connection to the suprarenal IVC (Fig 2) along with a large collateral circulation by the prevertebral plexus and azygos (Fig 3) No images suggestive of intra- or extravascular tumor or associated adenopathies were observed.

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Competition of interest: none.

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The lab results were normal, except for a slight increase in C-reactive protein and a D-dimer value of 15175 ng/ml (0-275). Markers of oncological, rheumatologic, or hypercoagulability disorders were negative. Serum protein electrophoresis and hormonal blood profiles were also normal. Anticoagulant treatment and compression with elastic stocking for both legs was started. The 3-month follow-up only showed mild residual edema of the right lower limb.

## DISCUSSION

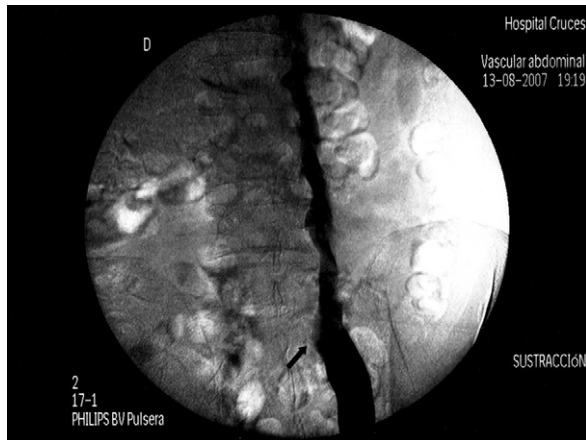
Iliac venous aneurysms are extremely rarely observed abnormalities. Following a systematic search in MEDLINE [1850-Nov 2007], EMBASE [1980-Nov 2007], and OVID with an unrestricted search strategy and with exploded MeSH terms (iliac veins/iliac venous, hypogastric, inferior cava vein, and aneurysms), only 21 reported articles were retrieved.<sup>1-21</sup>

In 1964, Abbot<sup>22</sup> classified these aneurysms as primary or secondary, according to their cause. The latter being considered a consequence of arteriovenous fistulas (AVF), proximal obstruction, or cardiovascular anomalies that increase the flow or pressure within the venous system.

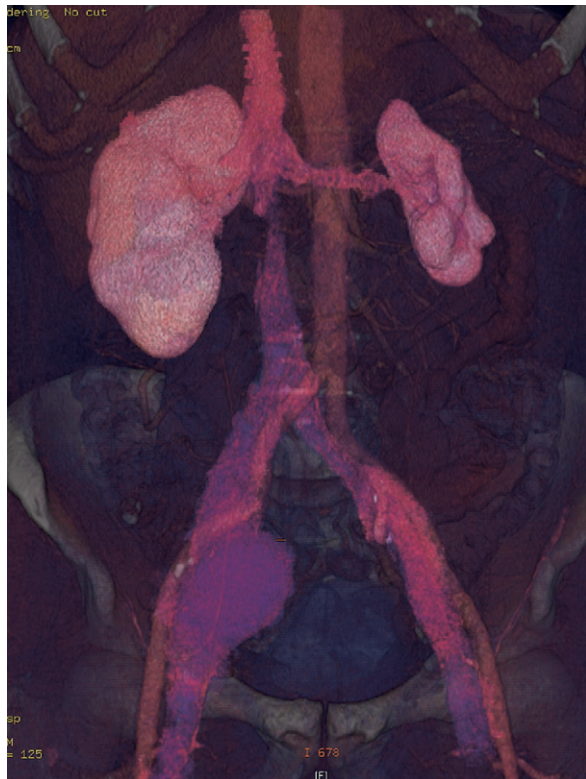
In our review, the AVFs, commonly due to prior trauma, were the most common cause (41%). This figure could be underestimated accounting that Mansfeld's review included at least 14 cases of aneurysms secondary to AVF between 1867 and 1984.<sup>5</sup>

The drainage impairment due to congenital anomalies represents 9% of our review. Our own case consisted of an anomaly due to hypoplasia of the infrarenal vena cava probably secondary to an atresia or failure in the embryological development of the junction between the hepatic segment and the right subcardial veins. The presence of a varicocele, a large collateral venous drainage throughout the prevertebral and azygos system, and the near complete flow obstruction of the IVC immediately below the renal veins support this diagnosis.

The iliac compression syndrome was clearly identified as the cause of the aneurysm secondary to flow obstruction in

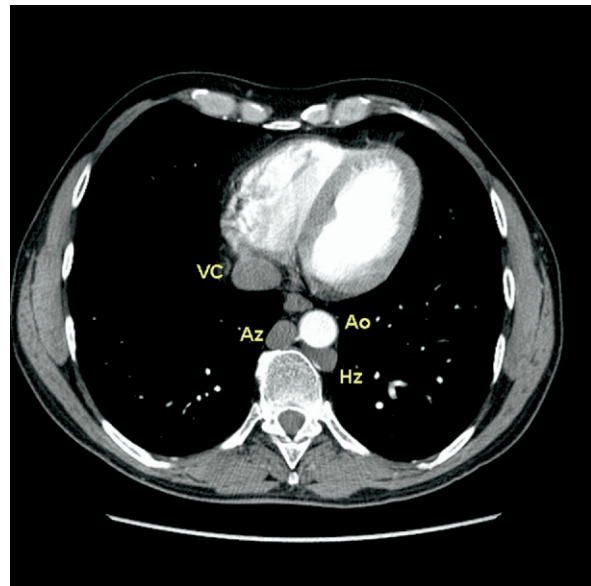


**Fig 1.** Ascending phlebogram showing complete occlusion of the left common iliac vein (arrow) and a large collateral circulation throughout the paravertebral plexus.



**Fig 2.** A 64-slice CT angiogram showing the iliac venous aneurysm and the hypoplasia of the infrarenal IVC.

only one case.<sup>8</sup> However, it is worth noting that in the overall analysis of the series, we found three cases corresponding to patients with high level of physical activity.<sup>8,9,15</sup> Hurwitz suggests that an obstruction of the venous return associated with an increased flow during exercise can generate sufficient venous hypertension to eventually dilate the vein over



**Fig 3.** CT scan showing the large collateral circulation throughout the azygos system. VC, Vena cava; AZ, azygos; AO, aorta; Hz, hemiazygos vein.

the time. Also of note, is the presence of three pregnant women in the review.<sup>4,13,20</sup> Uterine pressure, hyperperfusion, and hormonal changes related to gestation have been indicated as possible predisposing factors.<sup>4</sup>

Primary aneurysms are very infrequent with only seven reported cases.<sup>9,14,15,17,19-21</sup>

Finally, there is one case of venous aneurysm that developed after thermal lesion of the vein wall, which the author describes as the venous equivalent to an arterial pseudoaneurysm.<sup>16</sup> The most frequent clinical presentation among the reviewed cases was venous thrombosis (41%), followed by chronic venous insufficiency (32%). In our case, the patient presented with unilateral edema in spite of having a bilateral iliac thrombosis. We believe that this difference is due to the absence of thrombosis and preservation of the left femoropopliteal territory. Additionally, the large intrapelvic collateral circulation could also minimize the symptoms due to a common left iliac vein thrombosis.

Other initial symptoms previously reported have been abdominal pain or mass, hip pain, and pulmonary thromboembolism (PTE). Only four patients were diagnosed with absence of symptoms. Rupture of the aneurysm has also been described as a possible form of presentation.<sup>23</sup>

The analysis of different subgroups revealed that most aneurysms secondary to posttraumatic AVF were associated with symptoms of long-term chronic venous stasis (7 of 8). Patients with flow obstruction (congenital or acquired) remained asymptomatic until the onset of a thrombosis of the venous system. This is probably explained by the development of a compensatory collateral circulation over time. Similarly, patients with primary aneurysms did not present with long-term history of symptoms.

**Table I.** Review of the literature

	<i>Sex/age</i>	<i>Symptoms</i>	<i>Thromboembolic complication</i>	<i>Location</i>
Cornet (1969)	♂ 30 ♂ 50	Inflammatory injury and limb swelling Abdominal mass	None None	Right CIV Left EIV
Raitherl (1972)	♂ 48	Intermittent claudication and swelling	None	Left EIV
Vaccaro (1975)	♂ 65	Edema, leg ulcer and popliteal swelling	None	Right EIV
Parer (1984)	♀ 23	Asymptomatic	None	Left EIV
Mansfeld (1985)	♂ 56	Intermittent claudication and swelling	DVT lower limb	Right EIV
Valdes (1986)	♂ 58	Abdominal pain and constipation	None	Left hypogastric vein
Tisnado (1988)	♂ 57	Chronic venous stasis	None	Right EIV
Hurwitz (1989)	♂ 69	Painful limb swelling	Thrombosed aneurysm	Left CIV and EIV
Postma (1989)	♂ 33	Exercise intolerant + hemoptysis	Recent PTE	Left hypogastric vein
Gade (1991)	♂ 13	Limb swelling	Thrombosed right CFV	Left EIV and hypogastric junction
Salman (1994)	♂ 53	Chronic venous stasis, leg ulcer, and abdominal mass	Thrombosed distal SFV	Left CIV
Saito (1995)	♂ 19	Abdominal pain	Thrombosed IVC and left iliac veins	Right CIV and IVC junction
Labropoulos (1996)	♀ 34	Limb swelling	Thrombosed aneurysm	Right EIV and CFV
Petrunic (1997)	♂ 19	Abdominal pain	Thrombosed aneurysm	Right CIV
Alatri (1997)	♂ 39	Asymptomatic	None	Bilateral CIV
Jalaluddin (1998)	♀ 63	Pain in the hip and pulsatile swelling at right iliac fossa	None	Right EIV
Fourneau (1998)	♀ 21	Asymptomatic	None	Left EIV
Frikha (1999)	♂ 30	Chronic venous stasis and leg ulcer	None	Right EIV
Alonso (2002)	♂ 67	Limb swelling	Thrombosed aneurysm	Bilateral CIV
Banno (2004)	♀ 20	Asymptomatic	None	Left EIV
Cañibano (2007)	♂ 69	Back pain and limb swelling	Thrombosed aneurysm	Left CIV and EIV
Ysa (2008)	♂ 51	Fever and limb swelling	Thrombosed aneurysm	Right EIV

*CIV*, Common iliac vein; *EIV*, external iliac vein; *CFV*, common femoral vein; *SFV*, superficial femoral vein; *IVC*, inferior vena cava; *CFV*, common femoral artery; *SEA*, superficial femoral artery; *POP*, popliteal artery; *AVF*, arteriovenous fistula; *AVM*, arteriovenous malformation; *AR*, aneurysm resection; *DVT*, deep venous thrombosis; *PTE*, pulmonary thrombo embolism.

The use of ultrasounds, phlebogram, or computed tomography (CT) scan/angio magnetic resonance imaging (MRI) helps making the diagnosis in many occasions. In our case, the 64-slice angio-CT provided the most reliable information for diagnosis, showing a congenital anomaly of the vena cava, presence of a right venous aneurysm, and a thrombotic left iliac system probably secondary to the retrograde progression of the infrarenal vena cava thrombosis.

There appears to be a general agreement that abdominal and lower limb vein aneurysms should be corrected surgically once diagnosed, given their embologenic potential, the possibility of compression of adjacent structures, or even rupture.<sup>24,25</sup> There is also agreement over the need to re-establish venous continuity whenever possible to prevent the appearance of post-thrombotic syndrome.<sup>20</sup> When the flow is restored via bypass, a proximal AVF has also been used to prevent graft thrombosis.<sup>8</sup>

In our review, the surgical treatment was heterogeneous and frequently combined various techniques (Table I). Table II summarizes the treatments according to the cause of the aneurysm, the intrinsic characteristics of the group, and the permeability of the venous axis following the treatment. In the case of a diagnosed aneurysm following a symptomatic thrombosis, the latest recommendation seems to support medical treatment with anticoagulation and elastic compression measures.<sup>21</sup>

Although there are no reports of endovascular management of iliac venous aneurysms, posttraumatic and stenotic lesions of the ilio-caval segment have been corrected successfully with endoprosthesis.<sup>26</sup>

During an average follow-up of 9 months (15 days to 24 months), there was only one case of death secondary to a stroke and only one PTE. The most frequently reported complain was postphlebotic syndrome. In 32%

**Table I.** Continued

<i>Diameter (cm)</i>	<i>Related anomalies</i>	<i>Treatment</i>	<i>Outcome</i>	<i>Follow up</i>
Orange like	Posttraumatic AVF below adductor hiatus	Unsuccessful surgical review	Paretic post-thrombotic limb	12 mo
20	Posttraumatic AVF (SFA)	AVF ligation + vein ligation + subsequent AR (6m)	No complications	3 mo
No data	Posttraumatic AVF (SFA)	AVF ligation	No complications	17 d
25	Posttraumatic AVF (POP)	NONE	Death (stroke)	2 mo
10	Dialysis femoral-saphenous AV shunt	AVF ligation + anticoagulation	Aneurysm diameter reduction	6 wk
15	Posttraumatic AVF (SFA)	AVF ligation + subsequent AR	PTE + post-thrombotic limb	No data
10.6 × 8	Congenital AVM	Embolization + AR	Asymptomatic	12 mo
8 × 10	Posttraumatic AVF (SFA)	No data	No data	No data
8.8 × 4.4	Iliac compression syndrome	AR with graft reconstruction + AVF + anticoagulation	Thrombolysis of the graft 6m, thereafter patent	22 mo
3-4 approximately	None	Simple ligation at origin	Asymptomatic	12 mo
5 × 5	Congenital IVC hypoplasia	Unsuccessful thrombolysis + right femoral thrombectomy + AR	Asymptomatic	1.5 mo
25	Posttraumatic AVF (CFA) Iliac artery aneurysm	AVF ligation AR + end to end anastomosis Arterial aneurysm correction	Improvement of venous chronic insufficiency	9 mo
No data	Double IVC, hypoplastic right IVC, pre-renal stenosis of IVC	Anticoagulation	Asymptomatic	1 mo
No data	Double EIV estenosis	Vein ligation + anticoagulation	Post-thrombotic limb	24 mo
4.5 × 8.9	None	AR + lateral venorrhaphy + anticoagulation	Asymptomatic	12 mo
3.9 × 6.0	IVC, bilateral FEM-POP ectasia	None	No data	No data
4.3 × 7.3	Vein wall thermal injury	None	No complications	12 mo
3.5				
5 × 10	None	AR with contralateral SFV reconstruction + anticoagulation	Slight irritation saphenous nerve	18 mo
5 approximately	Posttraumatic AVF (SFA)	AVF ligation	No complications	No data
No data	5 cm IVC aneurysm	AR + anticoagulation	Post-thrombotic limb	6 mo
8	None	AR + lateral venorrhaphy + Anticoagulation	Asymptomatic	16 mo
11 × 4.2	None	Anticoagulation	No data	1 mo
5 × 9	Pre-renal IVC hypoplasia	Anticoagulation	Residual edema	3 mo

**Table II.** Summary of the different treatments according to the cause of the aneurysm and the permeability of the venous axis

	<i>n</i>	<i>Characteristics</i>	<i>AE without VFR</i>	<i>AE with VFR</i>	<i>No surgical treatment</i>
Primary (n = 7)	3	Thrombosed	1	1	1
	4	Nonthrombosed	0	3	1
Secondary (n = 15)					
Flow obstruction	4	Thrombosed	1	2	1
Thermal injury	1	Nonthrombosed	0	0	1
AVF <sup>a</sup>	7	AVF ligation	3	1	3
	3	No AVF ligation	0	0	3

AE, Aneurysm exclusion; VFR, venous flow reconstruction; AVF, arteriovenous fistula.  
<sup>a</sup>None thrombosed.

of cases, there was persistence or worsening of the previous symptomatology. Among the 15 patients in whom continuity of venous flow was preserved upon admission (postrevascularisation or absence of prior thrombosis),

only 7% showed residual edema of the lower limb. In the group where venous flow was not preserved, 50% of the patients showed residual edema (Fisher exact test,  $P = .053$ ).



This is the largest reported review in the literature with regard to iliac venous aneurysms. Despite the extensive descriptive analysis carried out, the low number of cases identified and the inter-group heterogeneity makes it difficult to draw possible conclusions with regard to its natural history and prognosis.

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