

# Pulmonary emboli due to venous aneurysm of extremities

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## Summary

Venous aneurysms are uncommon but they can have devastating consequences, including pulmonary embolism, other thromboembolic events and death. We report six cases of venous aneurysm of the extremities, in which the first sign of presence was acute pulmonary embolism. Surgical resection is recommended whenever possible. Our experience suggests that prophylactic surgery is cautiously recommended for low-risk patients with venous aneurysms of the abdomen and strongly recommended for extremity deep and superficial venous aneurysms for their potential risk of developing thromboembolic complications despite adequate anticoagulation. Other venous aneurysms should be excised only if they are symptomatic or enlarging.

**Key words:** Extremities vein aneurysm, pulmonary emboli, tangential venorrhaphy

## Zusammenfassung

*Pulmonalembolie verursacht durch ein venöses Aneurysma der Extremitäten*

Das venöse Aneurysma stellt eine sehr seltene Entität dar, mit jedoch potentiell verheerenden Folgen. Wir berichten über sechs Fälle von venösen Aneurysmen, die erst nach dem Auftreten einer Lungenembolie entdeckt wurden. Die chirurgische Sanierung wird, wenn möglich, empfohlen. Unsere Erfahrung zeigt, dass die chirurgische Prophylaxe bei low risk Patienten mit venösem Abdominalaneurysmen empfohlen werden kann; bei einem venösen Aneurysma der Extremitäten ist jedoch die chirurgische Intervention wegen des hohen Risikopotentials thromboembolischer Komplikationen selbst bei adäquater Antikoagulationstherapie absolut angezeigt. Andere venöse Aneurysmen sollten nur bei entsprechender Symptomatik entfernt werden.

## Introduction

Venous aneurysms (VAs) can be defined as a persistent isolated dilatation of twice or three times the normal diameter [15]. They have been reported in all major veins and are often misdiagnosed as soft tissue masses or as inguinal hernias. They can be divided into aneurysms of the deep and superficial system and are focal saccular or fusiform dilations that communicate with the accompanying normal vein via a single channel. Often they are of little clinical significance, but they can become a potentially life-threatening disease like deep-vein thrombosis with pulmonary embolism. VAs are classified according to their location and have been reported in association with the major veins of the neck, thorax, and abdomen as well as the deep and superficial veins of the upper or lower extremities [16–18]. VAs are usually asymptomatic and are detected incidentally on computed tomography (CT) or magnetic resonance (MRI) imaging scans [12].

Upper extremity VAs are usually asymptomatic and are most frequently treated because of their cosmetic appearance, while deep venous aneurysms have been reported to be associated with thromboembolism. And yet, the natural history of venous aneurysm remains poorly defined. In this study, we analyzed the venous system of six patients with venous aneurysms in their extremities and in which the first sign of their presence was acute pulmonary embolism. Our aim is to underline the importance of accurate diagnosis and surgical repair for preventing further embolic events.

## Case report 1

A 32-year-old woman presented with increasing respiratory distress and left thoracic pain without fever. D-dimer was altered (0.83 µg/mL, cut of value 0.4 µg/mL) by STA Liatest D-dimer (Diagnostica Stago, Asnières-Seine, France); in the absence of

other abnormal laboratory findings. The chest x-ray was normal, but an angio-thoraco-abdominal CT-scan revealed the presence of emboli in the segmentary branches of the left pulmonary artery with patency of the iliac-caval system. Venous Doppler ultrasound (US) examination of the extremities showed patency of the deep venous system of the lower extremities with a segmental isolated dilatation of the right great saphenous vein, measuring 3,1 cm (Fig. 1). An echogen area within the aneurysm was suggestive of a recent unstable thrombus. Under local anaesthesia and continued Valsalva manoeuvre the patient underwent resection of the mass. Histopathologic examination of the wall showed the characteristics of a venous aneurysm.

## Case report 2

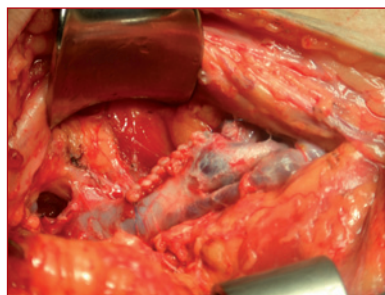
A 42-year-old man presented acute shortness of breath. There was no

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**Figure 1:** Venous duplex imaging demonstrating patency of the deep venous system with segmentary isolated dilatation of the great saphenous vein

history or physical finding consistent with venous thrombosis. The ventilation-perfusion scan demonstrated multiple segmental perfusion abnormalities consistent with and highly suggestive of pulmonary emboli. Venous Doppler US demonstrated the presence of a left saccular popliteal venous aneurysm 3.2 cm × 3.6 cm with normal vein above and below was. An evaluation for coagulopathy revealed the presence of circulating phospholipid antibodies (aPL). There was no history of previous venous thrombosis or trauma. The patient received anticoagulation therapy, and a temporary inferior vena cava filter (ICV filter) (Celect™ Vena CavaFilter) was inserted. The popliteal fossa was explored through a posterior approach and the patient underwent tangential aneurysmectomy and lateral venorrhaphy (Fig. 2). In the course of the operation the intraluminal thrombus was removed. The histopathologic examination revealed an aneurysm venous wall with focal reduplication of the internal elastic lamina.



**Figure 2:** Intraoperative image showing tangential aneurysmectomy and lateral venorrhaphy

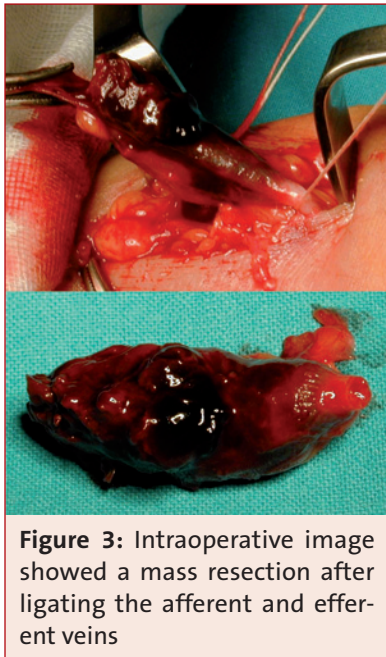
### Case report 3

A 43-year-old man in good health came to his local hospital with acute shortness of breath not associated with pleuritic chest pain or hemoptysis. His medical history was hypertension. The chest x-ray was normal, but a ventilation-perfusion scan demonstrated multiple segmental abnormalities highly suggestive of pulmonary embolism, venous Doppler US demonstrated the presence of a left thrombosed fusiform popliteal venous aneurysm 2.9 cm × 3.6. He was placed on anticoagulant therapy. The test for thrombophilia performed before the beginning of anticoagulant therapy was negative. Two hours later the patient experienced, in addition to acute shortness of breath, an associated, pleuritic pain and tachycardia with systolic hypotension (BP < 80 mmHg). A temporary ICV filter (Bard Recovery Cone® Removal System) was inserted. The patient subsequently underwent resection of the venous aneurysm with interposition of an autologous vein graft, a large amount of clot was found in the aneurysm. The histopathologic examination revealed an aneurysmic venous wall with thickened, fibrotic, moderately cellular intima adjacent to the densely fibrotic adventitia. There was a rarity of smooth muscle

### Case report 4

A 19-year-old man presented with increasing respiratory distress and right thoracic pain, in the absence of fever. His past medical history was negative for cardio-respiratory disease. The rest of the examination was unremarkable. Blood gases showed acute respiratory acidosis with a blood pH of 7.24, PaO<sub>2</sub> 81 mmHg, PaCO<sub>2</sub> 43 mmHg.

D-dimer was altered (0.42 µg/mL) in the absence of other abnormal laboratory findings. The chest X-ray was negative. Electrocardiography showed sinus tachycardia, while cardiac ultrasonography demonstrated mild right ventricular dilatation with minimal increase of the pulmonary pressures. The angio thoraco-abdominal CT-scan revealed the presence of embolism of the segmentary branches of the right pulmonary artery with patency of the iliac-caval system. The venous Doppler US scan showed bilateral patency of superficial and deep venous lower limb systems. The patient had noticed an asymptomatic soft mass in the right upper extremity, which had been present for about 3 years. Physical examination revealed an incompressible 2.9 × 0.5 cm mass in the lower third of the right arm. The duplex scan confirmed the diagnosis of the thrombosed fusiform venous aneurysm of the right cephalic vein. The venous aneurysm presented as a circumferential anechogenic area suggestive of a recent unstable thrombus. Under local anaesthesia the patient underwent resection of the mass, following ligation of the afferent and efferent veins (Fig. 3). The histology confirmed the characteristics of aneurysm, excluding the aspect of a varix.



**Figure 3:** Intraoperative image showed a mass resection after ligating the afferent and efferent veins

### Case report 5

A 46-year-old man presented with acute shortness of breath and varicose veins in the right leg. There was no history or physical finding consistent with venous thrombosis. D-dimer was altered (0.63 µg/mL) in the absence of other abnormal laboratory findings. The chest X-ray was negative. The angio thoraco-abdominal CT-scan revealed the presence of embolism to the segmentary branches of the left pulmonary artery with patency of the iliac-caval system. Venous Doppler US demonstrated reflux in the proximal and middle portions of the left small saphenous vein with perforator veins functioning normally. In the proximal part of the small saphenous vein there was a portion with a 2.6 cm aneurysm, thrombosed partially, whilst the bilateral deep vein system was patent. Under local anesthesia the patient underwent resection of the tender mass following ligation of the afferent and efferent limbs. The histologic examination confirmed the characteristics of a varix. Phleboc-

tomy of the remaining varices was performed.

### Case report 6

A 42-year-old man in good health experienced acute shortness of breath without pleuritic chest pain or hemoptysis. The chest x-ray was normal; but a ventilation-perfusion scan demonstrated multiple segmental perfusion abnormalities consistent with highly suggestive pulmonary emboli. Concomitant venous Doppler US demonstrated that the popliteal-femoral venous systems were patent bilaterally. Accurate evaluation of distal veins was not performed. No source of embolism was found. The etiology of PE remained unknown. The patient was placed on anticoagulant therapy and was discharged on acenocumarol and maintained prothrombin time (PT) at 2–3 INR. Tests for thrombophilia were negative. Three months later he again experienced acute shortness of breath associated with pleuritic chest pain and tachycardia; INR was 2.1; chest CT scan revealed bilateral filling deficits in the lungs suggestive of multiple pulmonary emboli. Again he had no clinical evidence of deep vein thrombosis, but an accurate venous Doppler US and subsequent CT scan (Fig. 4) revealed thrombosis of a 3.2 cm large posterior tibial vein aneurysm in the right leg with otherwise patent venous systems bilaterally. A temporary IVC filter (Celect™ Vena CavaFilter) was placed to prevent the risk of embolism during aneurysm repair. The patient subsequently underwent a tangential aneurysmectomy and lateral venorrhaphy; a large amount of clot was found in the aneurysm. The postoperative course was uneventful. The patient's demographics and characteristics are summarized in Table I.



**Figure 4:** CT scan revealed thrombosis of the right posterior tibial vein aneurysm

All patients had an uneventful recovery, and they were discharged on Acenocumarolo therapy for 3–4 months. In the three cases of deep vein aneurysm the temporary IVC filter was removed 3 months after discharge.

At 6–12 months follow-up, venous Doppler US examination demonstrated that the superficial and deep venous system was patent and free of venous reflux and symptoms in all cases.

### Discussion

Isolated venous aneurysm, without other varicosities is rare. VAs have been reported in the intra and extracranial veins, in the venous system of extremities, in the superior vena cava, the splenoportal and in common iliac veins.

Venous aneurysms are uncommon, to our knowledge the first characterization of venous aneurysm as a clinical entity was described 1915 by Osier during autopsy studies. The first symptomatic VA with pulmonary



Table 1: Patient demographic and characteristic data

Case	Sex	Age	Site	Medical history	D-dimer	Symptoms	Size, cm	Histology	Intervention
Case 1	F	32	great saphenous vein	none	0.83 µg/mL	increasing respiratory distress and left thoracic pain	3,1 x 2,9	aneurysm venous wall with endothelial denudation, attenuation of the elastic lamellae and medial fibrosis in areas of thrombus adherence	ligation/excision
Case 2	M	42	popliteal vein	circulating phospholipid antibodies (aPL)	0.94 µg/mL	acute shortness of breath	3,2 x 3,6	aneurysm venous wall characteristics with focal reduplication of the internal elastic lamina	tangential aneurysmectomy and lateral venorrhaphy
Case 3	M	43	popliteal vein	hypertension	1.05 µg/mL	acute shortness of breath not associated with pleuritic chest pain or hemoptysis	2,9 x 3,6	aneurysm venous wall with thickened, fibrotic, moderately cellular intima adjacent to a densely fibrotic adventitia and rare smooth muscle	resection of venous aneurysm with interposition autologous vein graft
Case 4	M	19	cephalic vein	none	0.42 µg/mL	increasing respiratory distress and right thoracic pain	2,9 x 3,5	aneurysm wall with fragmentation, and attenuation of the elastic lamellae, loss of smooth muscle cells,	ligation/excision
Case 5	M	46	small saphenous vein	leg varicose vein, CAD	0.63 µg/mL	acute shortness of breath	2,6 x 2,3	characteristics of varix	ligation/excision
Case 6	M	42	posterior tibial vein	none	0.69 µg/mL	acute shortness of breath	3,2 x 2,6	attenuation of the elastic lamellae, loss of smooth muscle cells	tangential aneurysmectomy and lateral venorrhaphy

CAD: Coronary artery disease

embolism was published by Dahl [5] in 1976. A soft tissue mass, changing in size according to the position of the limb or caused by the Valsalva maneuver suggest a venous aneurysm of the extremities. Asymptomatic accidental detection, local lower extremity symptoms or embolic pulmonary episodes may represent different aspects of manifestation of the same condition [13]. The aneurysms are usually asymptomatic, and the diagnosis is often casual or one of exclusion. Venous aneurysms in the extremities can be divided into aneurysms of the deep and superficial venous systems. The deep venous system seems to have a greater association with thromboembolism and more severe venous morbidity than the superficial system; however, our experience showed that even venous aneurysms of the superficial system can be a source of embolism. Pascarella et al. reported, for aneurysms of the superficial venous system, an incidence of 0,1 % [17]. The pathogenesis of venous aneurysm is not clear; several mechanisms have been suggested, such as reflux and venous hypertension, inflammation, infection, congenital vein wall weakness, mechanical trauma, hemodynamic, and localized degenerative change [20]. Histology and immunohistochemistry have shown a slackening of the venous wall in varicose veins and VA. Focal areas of endothelial denudation, fragmentation, and attenuation of the elastic lamellae, loss of smooth muscle cells, and medial fibrosis in areas of thrombus adherence were characteristic findings in VA and to a lesser extent in varicose vein specimens. The morphologic changes in both varicose veins and VAs may only involve only part of the circumference of the venous wall. The expression of matrix metalloproteinase was increased in venous aneurysms compared with normal vein

and varicose vein. Some authors suggest that this observation in conjunction with the morphologic changes in the extracellular matrix of venous aneurysmal lesions suggests a possible causal role for metalloproteinase in VA pathogenesis [14].

The asymmetric distribution of the disease process explains the feasibility of using lateral venorrhaphy to repair VAs.

To our knowledge venous aneurysms presented with pulmonary embolism in 24–32% or led to chronic venous disease in 76% [19]. Occasionally, superficial venous aneurysms can present with thromboembolism, but the incidence of pulmonary embolism due to superficial vein aneurysms is unknown; Gillespie et al. reported one case [11].

Rupture is a noted complication in arterial aneurysms but is rare in those of the venous system [7].

Diagnosis is usually performed by duplex scan following by CT scan which permit a correct diagnostic assessment of VA, or phlebography [2]. Venous Doppler US is the diagnostic method of choice for evaluation of venous aneurysms of extremities, since it is easy and quick and enables to define size and morphology of the aneurysm, however, we believe that before surgical repair a CT scan, MR angiography or ascending venography are mandatory for the assessment of deep veins and in order to delineate venous anatomy [2, 4].

Pulmonary embolic events represent the most frequent presentation of venous aneurysm. Risk is unpredictable and may be unrelated to the presence or absence of thrombus on imaging studies [1, 18]. Our experience and review of the literature suggest that anticoagulation therapy may be ineffective in preventing pulmonary embolism, therefore surgical repair is mandatory in all patients with symptomatic superficial or deep vein aneurysms and recommended

in asymptomatic patients. Repeated pulmonary embolism in patients with small asymptomatic isolated venous aneurysms without apparent evidence of thrombus on imaging studies require surgical treatment to avoid the development of pulmonary hypertension.

Pulmonary emboli with severe hemodynamic instability are in need of thrombolytic therapy to improve cardiopulmonary function and to reduce thrombus in the deep venous system aneurysm before the aneurysm repair [8].

ICV filter placement is useful before deep vein aneurysm surgical repair to reduce the risk of embolism during surgical aneurysm repair [8, 9]. Aneurysmectomy and lateral venorrhaphy is advised for saccular venous aneurysms, occasionally, they can be resected after prior ligation and in selected patients a graft can be placed. Fusiform aneurysms can be treated with resection and end-to-end anastomosis, resection with interposition graft, bypass or ligation of the proximal and distal vein. Superficial vein aneurysms can be treated by ligation of the afferent and efferent veins. Current endovenous ablation techniques are usually not feasible owing to aneurysm size and location [17]. Thus, treatment is primarily surgical and can be accomplished with simple ligation and excision [11].

After surgical repair we recommend anticoagulant therapy for three months [2].

To our knowledge anticoagulation may be ineffective in preventing pulmonary embolism during thrombosis of VA; early patency rates are encouraging, with no reports of recurrent pulmonary embolism following surgical repair, although the long term results of surgery are unknown [10], and only few cases of recurrence of VA after lateral tangential aneurysmectomy have previously been reported [6].

## Conclusions

Venous aneurysms are rare but they can often be potentially life-threatening, early diagnosis is necessary to prevent the risk of embolism or other major complications.

Our report underlines the importance of accurate evaluation of the venous system of the lower limb by duplex scan in all six cases of pulmonary embolism and the importance of approaching this disease entity on a case-by-case basis. Anticoagulation may be ineffective in preventing pulmonary embolism and early treatment by surgical repair is therefore recommended as a safe and effective procedure.

## Conflicts of interest

There are no conflicts of interest existing.

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Submitted: 08.11.2010

Accepted after revision: 20.05.2011