Hughes-Stovin Syndrome
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A 50-year-old woman suffering from shortness of breath and unproductive coughing for 6 months was admitted to the hospital because of recurrent hemoptysis. The patient had a history of deep venous thrombosis of both legs and the pelvic veins. A thrombophilic state was assumed because of a factor V Leiden mutation.

Angiography showed occlusion of both iliac vein systems and of the vena cava inferior (Figure 1). Venous access to the central circulation was possible through the superior caval vein. Although pulmonary angiography of the left lung showed some enlargement at the origin of the left upper-lobe pulmonary artery, right-sided pulmonary angiography revealed two large aneurysms originating from the right upper-lobe artery and the intermediate branch of the pulmonary artery (Figure 2).

The intermediate artery aneurysm was resected, and the artery was reconstructed using a saphenous vein graft. The patient also underwent right upper lobectomy because the right upper-lobe aneurysm reached too far into the periphery to allow reconstruction. Histological examination of the specimen showed vasculitis, and immunosuppressive therapy was started.

The Hughes-Stovin syndrome consists of deep venous thrombosis often involving the caval vein accompanied by single or multiple pulmonary arterial aneurysms. Massive pulmonary hemorrhage caused by rupture of an aneurysm is a frequent terminal event. Patients affected by this syndrome are often young adult men in the second to fourth decade of life. Case reports describing women with Hughes-Stovin syndrome are rare.

Figure 1. Injection of dye through a right femoral access shows occlusion of both the iliac veins and the inferior caval vein. Blood is drained by a collateral circulation of pelvic and lumbar veins (see Movie I).

Figure 2. Right-sided pulmonary angiography. Two large aneurysms originating from the upper lobe and intermediate pulmonary arteries. Marked hypoperfusion of the right upper and lower lobe is evident (see Movie II).
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