A 37-year-old man was seen for recurrent pulmonary embolism in 2007. He had previously experienced pulmonary embolism in 2001 and 2006. As a result, he developed significant pulmonary hypertension that manifested as shortness of breath on exertion. On physical examination, a large, rubbery, nonpulsatile mass of the right axilla (Figure 1) was noted. He was treated with therapeutic oral anticoagulation after his first pulmonary embolism, and an inferior vena cava filter was deployed after the second episode of embolism (in 2006). In spite of these measures, he experienced another episode of pulmonary embolism. A contrast chest computed tomographic scan (Figure 2) in 2007 revealed a large subcutaneous malformation in the right axilla and an acute large pulmonary embolus in the right-upper-lobe pulmonary artery. An ultrasound (Figure 3) of the axillary mass detected tortuous venous malformation with thrombus in situ that drained into the right axillary vein. The venous malformation was isolated from the venous circulation by deployment of 2 self-expanding stents (in 2007) in the right axillary and subclavian veins (Figure 4). He was continued on anticoagulation therapy and had no further embolic events through his last clinic visit at the end of 2009.

Chronic thromboembolic pulmonary hypertension due to recurrent pulmonary embolism from low-flow congenital venous malformations is reported in association with hypercoagulation. Clinical diagnosis is often delayed, and anticoagulation alone may not suffice to prevent pulmonary embolism. Management options for extremity lesions may include stripping of varicose veins or venous malformations, excision of vascular malformations, amputations, and debulking procedures. Because of the extensive and complex surgical procedure that is often required to treat the condition, percutaneous treatment may be considered to isolate the malformation from the venous circulation to prevent thromboembolism.

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Disclosures

None.
References


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