

**Spontaneous Hemothorax due  
to a Ruptured Pulmonary Artery  
Aneurysm in Rendu-Osler-Weber  
Disease**

**To the editor:** We read with interest the letter by Ausín and colleagues<sup>1</sup> reporting a case of spontaneous hemothorax due to rupture of a vascular malformation in a patient with Rendu-Osler-Weber disease. About half of pulmonary arteriovenous fistulas are found associated with this disease. Although spontaneous rupture of a malformation with massive hemothorax is an unusual event, the possibility of it happening is well known. The risk of rupture is

considerable in pregnant patients, in whom fistulas increase considerably in size.<sup>2</sup>

We treated a 24-year-old patient, in the 27th week of pregnancy, who had a family history of Rendu-Osler-Weber disease.<sup>3</sup> She was admitted with dyspnea and hypotension (70/40 mm Hg), tachycardia (120 beats/min), and tachypnea (40 breaths/min). Clinical manifestations and radiologic findings confirmed the presence of hemothorax. A pleural drain was inserted and 300 mL of blood was removed initially. Pulmonary arteriography detected an arteriovenous fistula. Because the patient's hemodynamic status was deteriorating and a large amount of blood had been lost through the pleural drain, emergency surgery was performed. A ruptured pulmonary arteriovenous fistula was found to be the source of massive bleeding. The fistula was sutured mechanically and resected. Additional fistulas were located, and one of the larger ones was also resected.

Congenital arteriovenous fistulas are usually asymptomatic but sometimes have nonspecific manifestations. The most common signs and symptoms are dyspnea, cyanosis, and polycythemia secondary to hypoxia caused by right-to-left shunt. Cerebral abscesses have also been described. Hemothorax can lead to severe bleeding and sudden death due to hypovolemic shock. Risk of this complication is reported to increase during pregnancy because of greater cardiac output and relaxation of smooth muscles induced by high concentrations of progesterone during gestation.<sup>4</sup>

This case illustrates a possible scenario that we believe should be borne in mind: when spontaneous hemothorax occurs in a woman of childbearing age, pregnancy and the presence of arteriovenous fistulas should always be ruled out. When a pregnant woman is known to have Rendu-Osler-Weber disease, it is important to warn of the risk of spontaneous hemothorax, which occurs most often in the second part of pregnancy.<sup>5</sup>

Another point we would like to make is that minimally invasive techniques can now be used to treat this complication. The combined use of selective embolization of a fistula and video-assisted thoracoscopy was recently described.<sup>6</sup> The patient must be hemodynamically stable before carrying out that procedure, however, and such was not the case for our patient. Pregnant patients with Rendu-Osler-Weber disease should be monitored carefully to assess increases in the size of arteriovenous fistulas. Prophylactic embolization or even surgical resection may be indicated in order to prevent massive hemothorax.<sup>5</sup>

**J. Freixinet, P. Rodríguez, and M. Hussein**

Unidad de Cirugía Torácica, Hospital,  
Universitario de Gran Canaria Dr. Negrín,  
Las Palmas de Gran Canaria, Spain.

1. Ausín P, Gómez-Caro A, Moradiellos FJ. Hemotórax espontáneo por rotura de aneurisma de arteria pulmonar en la enfermedad de Rendu-Osler-Weber. *Arch Bronconeumol.* 2004;40:602-3.
2. Gossage JR, Kani G. Pulmonary arteriovenous malformation: a state of the art review. *Am J Respir Crit Care Med.* 1998;158:643-60.
3. Freixinet J, Sánchez-Palacios M, Guerrero F, Rodríguez de Castro F, González D, López L, et al. Pulmonary arteriovenous fistula ruptured to pleural cavity in pregnancy. *Scand J Thor Cardiovasc Surg.* 1995;29:39-41.
4. Swinburne AJ. Pregnancy and pulmonary arteriovenous fistula. *N Y State J Med.* 1992;12:515-6.
5. Jakobi P, Weiner Z, Best L, Itskovitz-Eldor J. Hereditary hemorrhagic telangiectasia with pulmonary arteriovenous malformations. *Obstet Gynecol.* 2001;97:813-4.
6. Litzer PY, Douvrin F, Bouchart F, Tabley A, Lemercier E, Baste JM, et al. Combined endovascular and video-assisted thoracoscopic procedure for treatment of a ruptured pulmonary arteriovenous fistula: case report and review of the literature. *J Thorac Cardiovasc Surg.* 2003;126:1204-7.