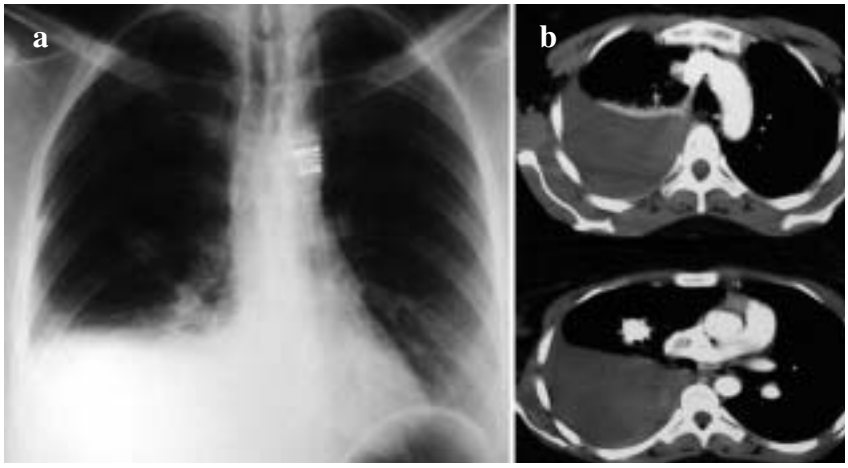

Figure 1. Chest x-ray showing right pleural effusion and increased density in the right parahilar region (a), and computed tomography of the chest showing an aneurysm of the right middle lobe artery and contrast in the right pleural cavity (b).

Spontaneous Hemothorax Due to Rupture of Pulmonary Artery Aneurysm in Rendu-Osler-Weber Syndrome

To the Editor: Pulmonary arteriovenous malformations appear in 2 to 3 per 100 000 population. Hereditary hemorrhagic telangiectasia, or Rendu-Osler-Weber syndrome, is a hereditary autosomal dominant disorder frequently associated with such malformations.[1] About 95% of these malformations are found in pulmonary circulation and only about 5% are systemic.[2] Spontaneous hemothorax is a rare manifestation of the disease.

We report the case of a 36-year old woman, smoker of 16 pack-years, with aspirin intolerance and an otherwise unremarkable history, except for the fact that her father had recently been diagnosed with Rendu-Osler-Weber syndrome. She came to the emergency department complaining of dyspnea and right-sided pleuritic pain developing over the course of only a few hours. Upon arrival she was pale, with signs of central cyanosis, tachypnea (28 breaths/min), and difficulty breathing evidenced by use of accessory muscles. Baseline oxygen saturation was 80%, blood pressure, 95/50 mm Hg, and temperature 36ºC. On lung auscultation, vesicular sounds were absent and the others were diminished in the lower half of the right hemithorax. Complete blood count, biochemistry, and coagulation tests were normal.

A chest x-ray showed right pleural effusion and increased density in the right parahilar region (Figure 1a). Contrast-enhanced computed tomography of the chest (Figure 1b) showed a vascular malformation arising from the right pulmonary artery and contrast extravasation from the malformation into the right pleural cavity.

Selective pulmonary arteriography showed a saccular dilation of the interlobar portion of the right middle lobe artery. This was interpreted as an aneurysmatic malformation that could not be embolized because of the high rate of blood flow in the artery. In view of the patient's hemodynamic instability and the characteristics of the aneurysm, an emergency thoracotomy was performed.

During surgery a hemothorax occupying nearly the entire chest cavity and an aneurysm of the interlobar portion of the right middle lobe artery were detected. The aneurysm was dissected and ligated, with atypical resection of the middle lobe without section of the venous branches.

The patient progressed satisfactorily during the postoperative period, with no further hemorrhages or complications, and was discharged on the seventh day following surgery.

Spontaneous hemothorax without pneumothorax is an uncommon entity of which hemorrhages due to pulmonary arteriovenous malformations are an unusual cause.[3] The most serious clinical manifestations of Rendu-Osler-Weber syndrome are paradoxical embolisms having repercussions on the central nervous system, massive hemoptysis, and hemothorax.[2,4] Arteriovenous malformations are usually diagnosed by computed tomography, and arteriography is a necessary diagnostic and therapeutic tool.[4] Currently, angiographic embolization techniques seem to be replacing surgery, as they are associated with lower rates of morbidity and mortality and allow lung function to be preserved.[1,5] In some patients (19%-60%), however, residual shunts persisting after embolization contribute to continued hypoxemia, thus rendering ineffective the attempt to preserve the parenchyma. Surgery remains the treatment of choice in large arteriovenous malformations requiring lung resection when embolization fails, when blood vessel rupture is suspected, when there is a high risk of hemorrhage due to the rupture of an aneurysm, and when there are recurrent hemorrhages.[1,6]

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