

# Unexplained Chronic Dyspnea in a Young Woman

R. Etter<sup>a</sup> G. Klammer<sup>a</sup> P. Probst<sup>b</sup> M. Tamm<sup>c</sup> F. Gambazzi<sup>d</sup> H. Borer<sup>a</sup>

Departments of <sup>a</sup>Internal Medicine and <sup>b</sup>Radiology, Bürgerspital Solothurn, Solothurn, and Divisions of <sup>c</sup>Pulmonary Medicine and <sup>d</sup>Thoracic Surgery, University Hospital Basel, Basel, Switzerland

A 32-year-old woman presented with exertional dyspnea (ED) 5 days after a knee arthroscopy. She suffered from poor exercise tolerance for years and from an increasing ED for months before the procedure. In 1986, a pulmonary shadow was found (fig. 1). On admission a systolic murmur was noticed in the left axillary line. Laboratory tests showed partial respiratory insufficiency [ $pO_2$  8.1 kPa (61 mm Hg);  $pCO_2$  4 kPa (30 mm Hg)] with normal values for D-dimer, cardiac and inflammatory parameters. During the 6-min walking test, oxygen saturation dropped from 89 to 82% and after the hyperoxic test  $pO_2$  was 18 kPa (135 mm Hg), indicating a shunt fraction of 26%. The chest CT revealed a pulmonary arteriovenous malformation (PAVM) in the left upper lobe (fig. 2, 3). Pulmonary embolism and deep venous thrombosis was ruled out by combined spiral CT, pulmonary angiography and CT phlebography. Therapy consisted of surgical resection of the anterior segment of the left upper lobe (fig. 4). Embolization therapy was considered to be unsuccessful because of the large feeding and draining vessels. The postoperative course was uneventful, ED disappeared rapidly and the shunt fraction decreased to 2%. No evidence of the Osler-Weber-Rendu syndrome was found.

PAVM is associated with the Osler-Weber-Rendu syndrome in up to 80%. The essential characteristic of PAVM is a right-to-left shunt. The most common presenting symptom is ED. The plain chest radiograph is usually

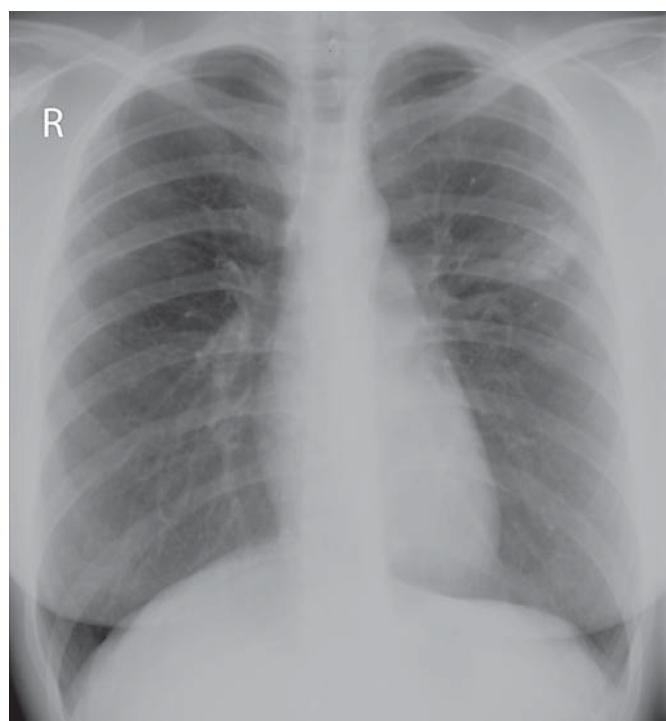
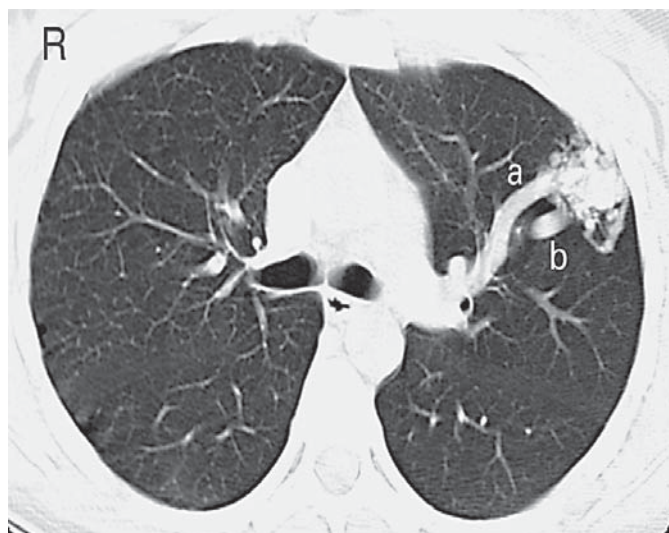
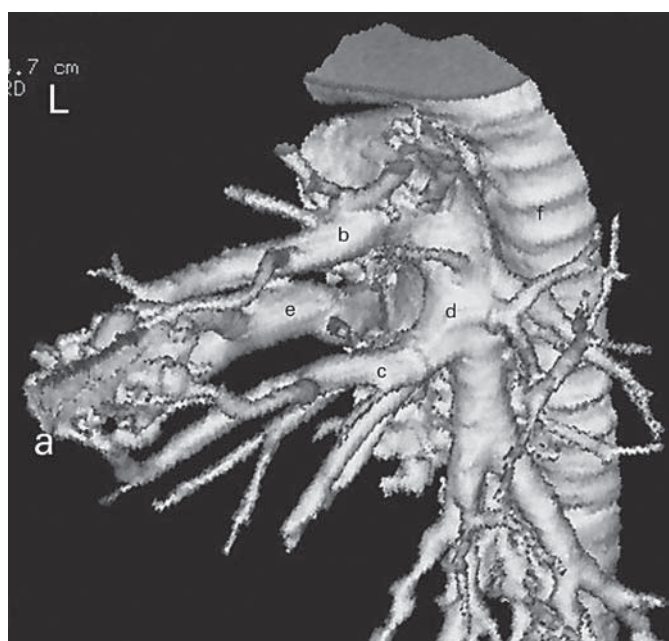


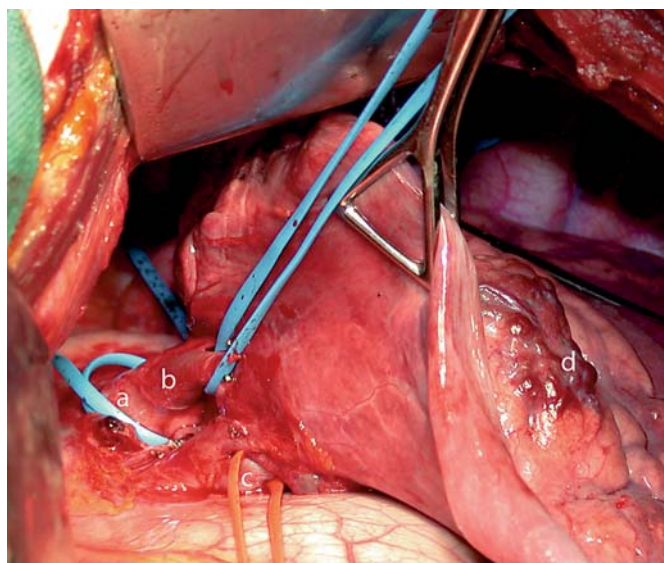
Fig. 1. Chest X-ray on admission: infiltrate in the left upper lobe.



**Fig. 2.** Chest CT on admission: PAVM in the left upper lobe with arterial feeding (a) and venous draining vessels (b).



**Fig. 3.** Three-dimensional reconstruction using chest CT, postero-lateral view: AVM (a), superior (b) and inferior arterial feeding vessels (c) arising from the pulmonary artery (d), big draining vein (e) and spine (f).



**Fig. 4.** Operation site: pulmonary artery (a), arterial feeding vessel (b), venous draining vessel (c) and AVM vessels on the lung surface (d).

abnormal. CT scan and pulmonary angiography are mostly diagnostic. PAVMs typically appear as peripheral, non-calcified lesions with a variety of vascular communications to the hilum [1, 2]. A rare complication is PAVM rupture with hemothorax [3]. The right-to-left shunt can be shown by contrast echocardiography. The shunt fraction is calculated using the hyperoxic test. Percutaneous embolization therapy is the treatment of choice for PAVM. Lung-sparing resection is an alternative procedure for symptomatic patients in whom embolization therapy was unsuccessful, technically not feasible (large PAVM vessels) or contraindicated.

## References

- 1 Gossage JR, Kanj G: Pulmonary arteriovenous malformations. A state of the art review. *Am J Respir Crit Care Med* 1998;158:643–661.
- 2 Khurshid I, Downie HG: Pulmonary arteriovenous malformation. *Postgrad Med J* 2002;78:191–197.
- 3 Varsano S, Edelstein E, Gendel B, Smorzik J: Bilateral and unilateral spontaneous massive hemothorax as a presenting manifestation of rare tumors. *Respiration* 2003;70:214–218.