Eur Respir J 2001; 17: 1328–1331
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European Respiratory Journal
ISSN 0903-1936

CASE FOR DIAGNOSIS

A 48-yr-old female with headache and dyspnoea

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Case history

A 48-yr-old white female was referred for an abnormality on her plain chest radiograph in the setting of progressive exertional dyspnoea. She had initially sought a medical evaluation 2 months previously for worsening headaches and a magnetic resonance imaging (MRI) scan of her brain was ordered. This MRI revealed an old infarct in her right posterior-inferior cerebellar hemisphere. She underwent a transoesophageal echocardiogram that showed a structurally normal heart but a positive bubble contrast study suggesting a right to left shunt. The cardiologist interpreting the study noted that there was no evidence of intracardiac shunt, but the delay after venous injection of the contrast bubbles until their appearance in the left heart suggested an anomalous, perhaps congenital, systemic venous connection to the left atrium. Two days after the echocardiogram was completed, the patient presented to a local emergency department complaining of chest discomfort. She denied any cough, fevers, night sweats, or weight loss. Past history was significant for ~20 pack-yrs of cigarette smoking, though she quit 15 yrs ago, and had 1–2 episodes of bronchitis per year.

The patient's physical examination showed: temperature 37.8°C; pulse 72 beats·min^{-1}; respirations 14 breaths·min^{-1}; and blood pressure (P) 135/75 mmHg. She was mildly obese and in no distress. Examination of skin, nose and mouth revealed no abnormalities. Examination of the neck revealed no elevation in jugular venous pressure. Chest examination revealed symmetric breath sounds with no crackles, wheezing or rhonchi and resonance to percussion bilaterally. Cardiac examination revealed a regular rhythm with normal first and second heart sounds and no murmurs or pericardial friction rub. The abdomen was soft, nontender and without hepatosplenomegaly. There was no cyanosis, clubbing or oedema. The neurological examination was normal.

The laboratory findings were: total leukocyte count 7.54×10^9 L^{-1} with 43% neutrophils, 44% lymphocytes and 8% monocytes, hematocrit of 39.7% and platelet count of 286×10^9 L^{-1}. Serum chemistries, blood urea nitrogen (UN) and creatinine were all normal. An arterial blood gas obtained in room air with the patient sitting revealed a pH of 7.43, an oxygen tension (P_{a,O_2}) of 9.7 kPa and a carbon dioxide tension (P_{a,CO_2}) of 5.6 kPa.

A posterior-anterior chest radiograph was taken (fig. 1) and a computed tomography (CT) scan of the thorax was obtained (fig. 2).

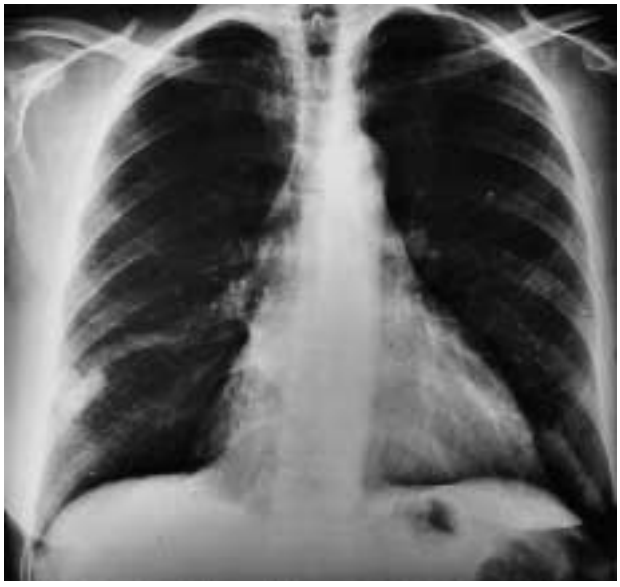

Fig. 1.–Plain chest radiograph showing an abnormality in the right lung.

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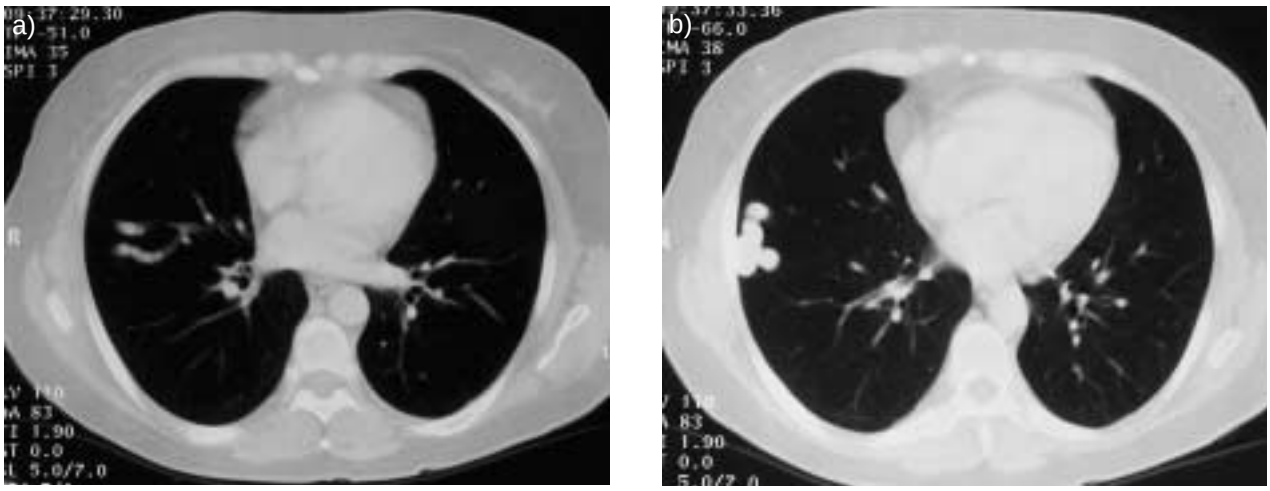

Fig. 2.–Thoracic computed tomography scan revealing proximal a) and distal b) views of the abnormality in the right lung.

BEFORE TURNING THE PAGE, INTERPRET THE PLAIN CHEST RADIOGRAPH AND THORACIC CT SCAN AND SUGGEST A DIAGNOSIS

Interpretation

Chest radiography

An ~2 cm lobulated nodule can be seen in the right lung base on the posterior-anterior view (fig. 1) with a smooth tubular structure suggesting a vascular connection.

Computed tomography

CT scanning demonstrated a pair of contrast-enhanced vascular structures (fig. 2a) connecting the right hilum to the 2.5 cm nodule in the anterolateral aspect of the right lower lobe (fig. 2b). This pattern of vascular structures in the present clinical setting made pulmonary arteriovenous malformation the most likely diagnosis.

Angiography

Selective angiogram of the right lower lobe demonstrated a 6.1 mm arterial feeding vessel with direct connection to and contrast enhancement of a similar size venous draining vessel (fig. 3).

Diagnosis: "Pulmonary arteriovenous malformation"

Clinical course

The patient in this report had a 6.1 mm arterial feeding vessel on angiography that was successfully occluded with three metal coils (fig. 4). No other arteriovenous malformations were detected on pulmonary angiography. Several months after the embolization procedure, the patient had experienced no further headaches and reported a marked improvement in her exertional dyspnoea (repeat resting arterial blood gas testing was not performed). Her primary physician will monitor her clinically for any further symptoms suggestive of pulmonary arteriovenous malformations (PAVMs) and within 1 year, a pulse oximetry evaluation and a follow-up chest radiograph were recommended to determine if the feeding artery and vein had collapsed.

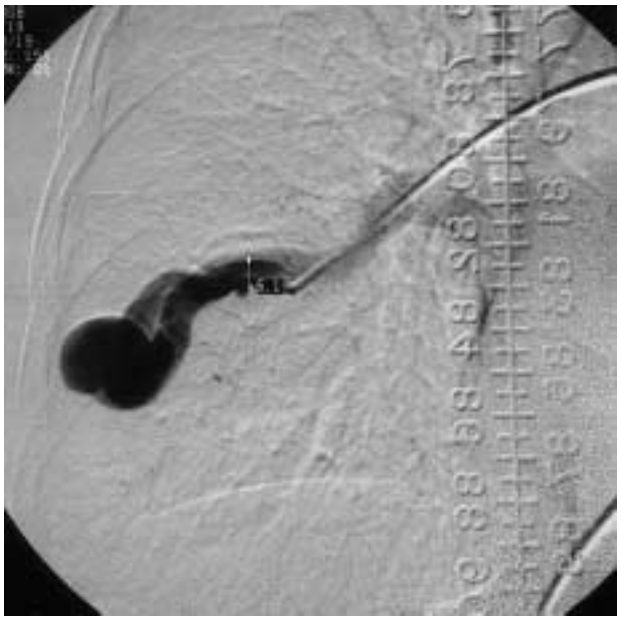

Fig. 3.–Fluoroscopic image during selective angiography demonstrating the calibre of the arterial feeding vessel and the pulmonary arteriovenous malformations.

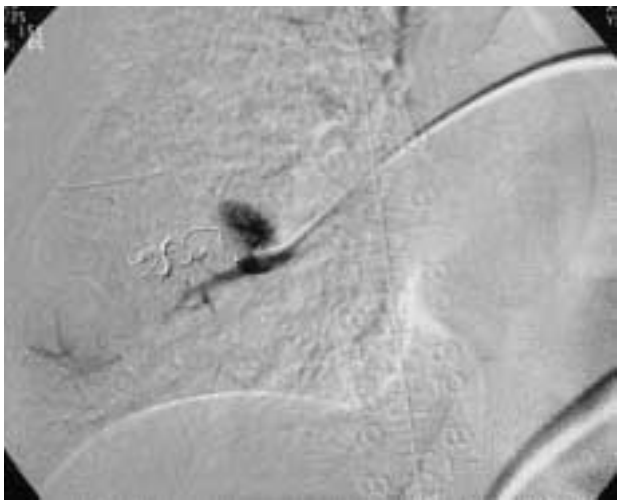

Fig. 4.–Fluoroscopic image after successful embolization with visible metal coils and without pulmonary arteriovenous malformations filling during selective angiography.

Discussion

PAVMs are defined as abnormal pulmonary vascular communications comprised of a supplying arterial source connected to a draining venous source, either by a persistent foetal anastomotic channel or acquired, disease-induced communications. Equivalent terms include pulmonary arteriovenous fistulae, pulmonary arteriovenous aneurysms, pulmonary haemangiomas and pulmonary telangiectasias [1]. The first reported case of PAVM was described at autopsy in 1897 by CHURTON [2]. The prevalence of PAVM has been estimated in the general population as 1:39,216 in northern England, 1:16,500 in the USA and 1:3,500 in Denmark.

Approximately 70% of cases of PAVM are associated with hereditary haemorrhagic telangiectasia (also HHT or Osler-Weber-Rendu Disease), though the patient was diagnosed with isolated congenital or sporadic PAVM as she had no evidence or family history of HHT.

Complications of PAVMs include haemoptysis and compromise of the lung's ability to exchange oxygen and filter the blood resulting in hypoxaemia, paradoxical embolization, neurological symptoms (such as headache) and brain abscess; the first three of these occurred in the patient. As many as 71% of patients

with PAVMs secondary to HHT experience headache symptoms and ~55% satisfy criteria for the diagnosis of migraine [3]. Though frequency of headache is unknown for patients with sporadic PAVMs, the presentation of the patient with headache, which resolved after PAVM embolization, suggests that the aetiology of her headache was related to the PAVM.

The decision to treat sporadic PAVMs is generally based on a history of associated complications and/or the risk for complications based on PAVM size. WHITE *et al.* [4] proposed that all PAVMs with feeding arteries ≥3 mm be occluded. Their experience over a 6-yr period included treating all patients with a feeding vessel >3 mm, which resulted in no further cerebrovascular events as well as stabilization of P_{a,O_2} values [4]. Reported mortality from untreated PAVMs ranges 0–29% [5], though there have been no prospective trials to assess the relative benefit of observation *versus* treatment.

Treatment until 1980 was limited to surgical resection and ligation, but improved catheterization techniques have made embolotherapy the treatment of choice for most patients [6]. Embolization is often accomplished with detachable balloons, metal coils or a combination of both. The advantages of embolization include the ability to treat patients with multiple pulmonary arteriovenous malformations, as often occurs in hereditary haemorrhagic telangiectasia, and those who are unable to tolerate surgery. Potential major complications of embolotherapy include systemic embolization of deflated balloon or coils and pulmonary infarction if other pulmonary artery branches are inadvertently occluded.

References

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