Surgery for multiple and diffuse pulmonary arteriovenous fistulas during childhood

Makoto Takahama, MD, PhD
Ryoji Yamamoto, MD, PhD · Ryu Nakajima, MD, PhD
Nobuhiro Izumi, MD, PhD · Hirohito Tada, MD, PhD

Abstract

We report an uncommon clinical case of multiple, diffuse pulmonary arteriovenous fistulas that occurred during childhood. A 15-year-old Japanese girl with multiple pulmonary arteriovenous fistulas presented with moderate hypoxemia and was treated by left lower lobectomy for complete resection of the arteriovenous fistulas. Clinical and radiographic features are presented.

Key words
Vascular disease · Fistula · Shunts · Lung, congenital lesions

Introduction

Pulmonary arteriovenous fistulas (PAVF) are rare congenital malformations that result from errant capillary development, with incomplete formation or disintegration of the vascular septa that normally divide the primitive connections between venous and arterial plexuses.1 PAVFs are characterized by a right-to-left shunt, which reduces arterial oxygen saturation. The effect of these connections depends on the size of the vessels involved.2 Almost all PAVFs are the single type; diffuse, multiple-type PAVFs are rare. We describe a case of multiple, diffuse PAVFs that was treated by left lower lobectomy.

Case

A 15-year-old girl was referred with cyanosis, clubbing of the fingers, and dyspnea on exertion, which had first been noticed at 14 years of age. Abnormal shadowing had been noted on chest radiography since she was 9 years old, with no medical follow-up. Physical examination revealed no continuous murmur and no abnormal lung sounds. The family history was noncontributory, and no relation to hereditary hemorrhagic telangiectasia was identified.

Contrast-enhanced chest computed tomography (CT) revealed PAVFs scattered profusely throughout the left lower lobe accompanied by dilatation of the left inferior pulmonary vein and inferior phrenic veins (Fig. 1). Pulmonary arteriography showed abnormal diffuse vessels after contrast medium filled the arterial branch, and prompt venous return through PAVFs was clearly apparent. Quantitative perfusion scintigraphy and whole-body imaging using 99mTc-macroaggregated albumin (MAA) documented an estimated perfusion rate of 13% in the left lung (Fig. 2a) and radioactive accumulation in the lungs, kidneys, liver, spleen, thyroid, and brain, respectively (Fig. 2b). This accumulation of MAA was consistent with the presence of right-to-left shunt, and the calculated shunt value was 42.9%.3 Electrocardiography revealed moderate left ventricular hypertrophy due to right-to-left shunt. Peripheral blood cell analysis showed moderate polycythemia (red blood cell count 6.19 × 10^12/l; hemoglobin 14.4 g/dl; hematocrit 45.6%), attributed to chronically sustained hypoxemia. Chemical and serological analyses of the blood showed no other abnormalities. Arterial blood gas analysis with the patient supine position inhaling room air revealed moderate hypoxemia with hypocapnia (pH 7.420; PaO_2 54.1 mmHg; PaCO_2 33.4 mmHg).
Fig. 1  

a Preoperative contrast-enhanced computed tomography (CT) demonstrates pulmonary arteriovenous fistulas (PAVFs) scattered profusely throughout the left lower lobe but are absent in the other pulmonary lobes.  

b Three-dimensional CT reveals multiple feeding arteries and multiple drainage veins in the left lower lobe.

Fig. 2  

Technetium-99m-macroaggregated albumin (99mTc-MAA) posterior imaging documented an estimated perfusion rate of 13% in the left lung (a) and radioactive accumulation in the lungs, kidneys, liver, spleen, thyroid, and brain (b).