Pulmonary Arteriovenous Fistula Manifesting as Amaurosis Fugax
—Case Report—

Munekazu YAMAKUCHI, Shigeya TANAKA*, Tetsuzo TOMOSUGI*, Koichi MOROKI*, Masahiko YAMADA*, Hideshi TOUJOU*, Koichi UETSUHARA*, and Ikuro MARUYAMA

Department of Molecular Laboratory Medicine, Kagoshima University School of Medicine, Kagoshima; *Department of Neurosurgery, Kagoshima City Hospital, Kagoshima

Abstract

Pulmonary arteriovenous fistula (PAVF) is a rare condition which occasionally causes neurological complications. A 43-year-old female with multiple PAVFs presented with several episodes of amaurosis fugax and transient right hemiparesis. She had no other vascular abnormality, and her human leucocyte antigen haplotype did not coincide with previous patients with hereditary hemorrhagic telangiectasia. She underwent PAVF embolization to prevent further neurological complications, and had an uneventful subsequent clinical course. Amaurosis fugax is a slight neurological symptom and may be an early important sign of PAVF. We stress that PAVFs should be considered in the differential diagnosis of patients with amaurosis fugax who complain of exertional dyspnea or demonstrate cyanosis.

Key words: pulmonary arteriovenous fistula, amaurosis fugax

Introduction

Pulmonary arteriovenous fistula (PAVF) was first described in 1897.29 PAVF may appear as an isolated anomaly or with hereditary hemorrhagic telangiectasia (HHT) (Rendu-Osler-Weber disease), which is a bleeding disorder transmitted as an autosomal dominant trait.1,14,18 Approximately 30% of patients with PAVF have associated HHT, and 18% without HHT develop neurological complications.17,18 Initial clinical manifestations include thrombotic or embolic stroke,10 brain abscess,12,20 and transient ischemic attacks (TIAs).4,29 The clinical triad of cyanosis, exertional dyspnea, and digital clubbing is common, but many patients are asymptomatic at diagnosis. Consequently, some patients refuse treatment for the fistula. However, the clinical course is thought to be far from benign, because the neurological complications, although rare, are potentially disastrous.8,29 We describe a patient who suffered amaurosis fugax and transient hemiparesis resulting from multiple PAVFs.

Case Report

A 43-year-old female was admitted to our hospital in July 1994 with sudden onset of right hemiparesis. She had undergone a routine health examination at the age of 12 years. Chest radiography had disclosed a lung mass. Pulmonary angiography revealed a PVAF. She had suffered exertional dyspnea and cyanosis involving the lips since 20 years of age. Surgical excision of the PAVF was recommended, but she refused treatment. Since May 1994, she has experienced three episodes of amaurosis fugax of the right eye which lasted for a few seconds.

On admission, she was afebrile, alert, and well-oriented. She had slight right hemiparesis and gait disturbance, but no cranial nerve paresis. Physical examination revealed marked clubbing of her fingers, but no cutaneous telangiectases suggestive of Rendu-Osler-Weber disease. Her heart rate was regular at 70 beats/min and her blood pressure was 108/60 mmHg. A continuous bruit was audible at the second intercostal space on the right which increased with inspiration.

Her optic disks showed no telangiectases or stenotic vascular changes. Laboratory data included...
a hemoglobin concentration of 17.1 g/dl and a hematocrit of 51.0%, indicative of polycythemia due to chronic hypoxemia. Routine blood biochemistry tests were normal. Arterial blood gas analysis revealed severe hypoxemia with PaO₂ of 61.2 mmHg, PaCO₂ of 34.3 mmHg, and SaO₂ of 92.4% breathing room air. These data were compatible with a right-to-left shunt due to PAVFs. Chest radiography on admission showed a well-defined ovoid mass in the right upper lobe (Fig. 1). Skull radiography, computed tomography of the brain with and without contrast medium, magnetic resonance imaging of the brain and spinal cord, angiography of the aortic arch and both common carotid and vertebral arteries, and electroencephalography disclosed no abnormal findings (Fig. 2). Electrocardiography and echocardiography

Fig. 1 Chest radiograph on admission showing a mass in the right upper lobe of the lung.

Fig. 2 Anteroposterior views of the right (upper left) and the left (upper right) internal carotid angiograms, and lateral views of the right carotid bifurcation (lower left) and aortic arch (lower right) demonstrating no abnormalities.

Fig. 3 Pulmonary arteriogram before embolization showing multiple bilateral pulmonary arteriovenous fistulas (PAVF): two in the right upper lobe (left) and one in the left upper lobe (center). The three PAVFs had completely disappeared after embolization (right).
also demonstrated no intracardiac or valvular lesions. Pulmonary arteriography revealed multiple bilateral PAVFs: two in the right upper lobe and one in the left upper lobe (Fig. 3 left, center). Her human leukocyte antigen (HLA) haplotype was HLA-A2A24B37B52Cw6. Eventually, coil embolization was recommended, though all neurological signs had disappeared within 24 hours of admission.

In October 1994, she underwent embolization of the three PAVFs after informed consent was obtained. Afterwards, she suffered no further TIAs (Fig. 3 right). Three months later, her SaO2 rose from 92.4% to 96.4%, other blood gas analysis results normalized, and her hematocrit decreased to 41.7%. No complications or exertional dyspnea have become evident during 5 years of follow up.

**Discussion**

The severity of neurological complications depends on the degree of right-to-left shunting.\(^1,8,14,18,20\) The major causative mechanisms are polycythemia due to hypoxemia\(^9\) and paradoxical emboli.\(^10,16\) Polycythemia is known to increase the possibility of cerebral thrombosis, due to the high viscosity of the blood and reduction in cerebral blood flow.\(^9\) However, most patients with PAVF develop neurological complications secondary to the formation of embolic materials within the PAVF (termed “paradoxical emboli”) that pass into the arterial circulation through the pulmonary vein. These emboli, once transmitted into the intracerebral circulation, can cause neurological complications ranging from mild sensory and/or motor disturbances to severe hemiplegia or brain abscess. Most reported neurological complications due to PAVF have involved permanent damage such as stroke or brain abscess. Cerebral abscesses occurred in 9%, strokes in 18%, and TIAs in 37% of patients with PAVF, and strokes are likely to be preceded by TIAs.\(^20\) Identification of TIAs heralding the onset of strokes is important.\(^1,8,20\)

Accordingly, the diagnosis of PAVF can be made prior to the occurrence of an irreversible neurological deficit by careful observation of patients with TIAs.

The accepted treatment for PAVF is resection when dealing with isolated lesions, but therapeutic embolization is preferable for multiple PAVFs.\(^10,21\) Since multiple PAVFs were found in our patient, coil embolization was performed with good results. Transient hemiparesis prompted the PAVF embolization, since she had been untreated for 20 years.

Interestingly, recurrent amaurosis fugax involving the right eye was the first presenting symptom in our patient. The visual disturbances consisted of sudden onset of partial or complete loss of monocular vision that lasted for seconds to minutes, followed by complete recovery. Generally, amaurosis fugax can be caused by emboli passing into the ophthalmic circulation from the ipsilateral common carotid artery and its branches.\(^6,7\) However, there are many other causes such as cardiogenic emboli,\(^21\) retinal vascular insufficiency,\(^46\) collagen vascular diseases,\(^5\) migraines,\(^13\) drugs,\(^13\) vasospasm,\(^20\) and local disk anomalies. These etiologies were considered unlikely in our patient on the basis of various examinations. Although previously unknown, amaurosis fugax in our patient was probably caused by paradoxical emboli from the PAVFs, since the amaurosis fugax completely resolved after treatment. Furthermore, the different onset of symptoms and multiple localities of the lesion strongly suggest an embolic mechanism.

Multiple PAVFs are likely to occur in patients with HHT.\(^17\) PAVFs linked with HHT occasionally enlarge and/or cause neurological complications.\(^16\) In our patient, although no vascular anomaly except for the PAVFs was detected by multiple imaging studies, PAVFs could be a manifestation of HHT. HLA-A2, Bw17, and Bw35 are all related to HHT in patients with PAVFs.\(^11\) The HLA haplotype in our patient was HLA-A2A24B37B52Cw6, which did not coincide with previous reports except for HLA-A2. Thus, further research regarding the relationship between HLA types and clinical manifestations of PAVF is necessary.

The present patient with PAVFs and associated neurological complications, amaurosis fugax, and transient hemiparesis was successfully treated by PAVF embolization. The combination of these neurological symptoms, exertional dyspnea, and cyanosis should prompt the consideration of PAVF in the differential diagnosis. If the patient is conscious of amaurosis fugax, embolization of the fistula is recommended.

**References**


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Address reprint requests to: M. Yamakuchi, M.D., Department of Molecular Laboratory Medicine, Kagoshima University School of Medicine, 8–35–1 Sakuragaoka, Kagoshima 890-8520, Japan.