Cerebellar Abscess Associated with Pulmonary Arteriovenous Fistula and Hereditary Hemorrhagic Telangiectasia

—Case Report—

Masahiro OGINO^{*,**}, Hiroshi INOUE^{***}, Shun-ichi HARADA^{**},
Hirohisa HORINOUCI[†], and Tsuneo NAKAMURA^{**}

*Departments of *Neurosurgery and †Surgery, School of Medicine, Keio University, Tokyo; **Department of Neurosurgery, Saiseikai Kanagawa-ken Hospital, Yokohama; ***Department of Neurosurgery, Tokyo Dental College Ichikawa Hospital, Ichikawa, Chiba*

Abstract

A 57-year-old male with a past history of bilateral pulmonary arteriovenous fistulas (PAVFs) experienced a sudden onset of headache and gait disturbance. There was a family history of PAVF and recurrent epistaxis. He had diffuse telangiectasia of the tongue, and hereditary hemorrhagic telangiectasia was diagnosed. Neuroimaging revealed a brain abscess in the right cerebellar hemisphere, which was successfully aspirated under ultrasound guidance. The PAVFs were resected afterwards in two-staged operation. No recurrence of the abscess has been observed. Cerebral abscesses complicated by PAVF are usually supratentorial. Complete eradication of PAVF is essential because the brain abscess will sometimes recur if the PAVF is left untreated.

Key words: cerebellar abscess, hereditary hemorrhagic telangiectasia (Rendu-Osler-Weber disease), pulmonary arteriovenous fistula

Introduction

Hereditary hemorrhagic telangiectasia (HHT) or Rendu-Osler-Weber disease is an autosomal dominant disorder which is characterized by multiple telangiectases of mucocutaneous tissues and various organs. The classical triad of HHT, familial occurrence, multiple telangiectasia, and recurrent epistaxis, is not always discovered in patients, and cutaneous telangiectases are sometimes misdiagnosed as moles or freckles in youth.⁷⁾ Pulmonary arteriovenous fistula (PAVF) is present in about one fifth of patients with this disease,^{2,11)} and 50–60% of the patients with PAVF also have HHT.¹⁹⁾ Neurological complications are observed in 29–33% of the patients with PAVF,^{15,19)} and 8–12% of the familial

members with HHT.^{5,11)} The incidence of neurological disorders in patients with PAVF and HHT is 41%. Although 61% of the neurological disorders of HHT are secondary to concomitant PAVF, HHT is associated with central nervous system lesions such as vascular malformations of the brain and spinal cord (36%) and portal-systemic encephalopathy (3%).¹⁹⁾

About 5% of the patients with PAVF have cerebral abscesses¹⁹⁾ due to the following: 1) hypoxemia decreases resistance of cerebral tissue to infection; 2) arterial thrombosis due to secondary polycythemia causes cerebral ischemia/infarct which is optimal to bacterial growth; and 3) septic embolus through PAVF.¹³⁾ Cerebral abscess occurs in 13% of the patients with PAVF and HHT,¹⁹⁾ but in only 3–

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Author's present address: M. Ogino, M.D., Department of Neurosurgery, Saiseikai Utsunomiya Hospital, Utsunomiya, Tochigi, Japan.

5% of patients with congenital heart disease.²³⁾ Therefore, HHT with PAVF has a relatively high association with cerebral abscess, usually in a supratentorial location, possibly due to associated unrecognized disorders such as angiographically occult cerebral telangiectasia.

We describe a case of cerebellar abscess secondary to HHT and PAVF.

Case Report

A 57-year-old businessman was admitted to our hospital because of sudden headache on November 28, 1990. He had an episode of recurrent epistaxis in his youth. Routine examination had indicated a PAVF in 1975. He had no complaint at the time and received no further examination or treatment. In April 1989, he was admitted to our hospital because of pneumonia and sepsis. Pulmonary angiography revealed bilateral PAVFs with 27% shunt rate. He declined treatment, and was discharged but followed up at the surgical outpatient department. Family history revealed PAVF in his mother and recurrent epistaxis in his younger brother and his daughter.

Physical examination disclosed clubbed fingers with peripheral cyanosis and telangiectasia of the tongue (Fig. 1). A continuous murmur accentuated in the systolic phase was heard in the right lung field. He was alert and without neurological symptoms except for mild gait disturbance. No marked cerebellar sign was observed. There was no abnormality in his optic fundi.

Blood tests revealed that his hemoglobin level was 16.4 g/100 ml, hematocrit value 48.9%, and white blood cell count 6300/mm³. Arterial blood gas measurements in room air were PO₂ 76.8 mmHg, PCO₂ 37.7 mmHg, and pH 7.42. Chest radiography

Fig. 1 Photograph showing telangiectasia on the tongue.

Fig. 2 Radiographs of the PAVFs showing the noncalcified nodular lesions (arrowheads) on the chest x-ray film (left) are well visualized on the right pulmonary angiogram (right).

Fig. 3 Precontrast CT scan (left) on admission demonstrating a low density lesion in the right posterior fossa with postcontrast ring enhancement (right).

showed noncalcified nodular lesions in the right lower lobe with vascular markings to the hilum (Fig. 2 left). Pulmonary arteriography performed at the previous hospitalization had showed one PAVF in the right upper lobe, as well as one in the lower lobe and two in the middle (Fig. 2 right).

Precontrast computed tomography (CT) demonstrated a low density lesion in the posterior fossa with postcontrast ring enhancement (Fig. 3). Magnetic resonance (MR) imaging with gadolinium-diethylenetriaminepenta-acetic acid (Gd-DTPA) revealed a ring-enhanced mass in the right cerebellar hemisphere just beneath the tentorium. The T₂-weighted image showed a hypointense area surrounded by perifocal edema (Fig. 4). Vertebral angiography showed an avascular area in the right posterior fossa but no vascular abnormality (Fig. 5).

The abscess was drained under ultrasound

Fig. 4 MR images on admission (A: axial proton-weighted image, B: axial T₂-weighted image, C: axial T₁-weighted image with Gd-DTPA, D: sagittal T₁-weighted image with Gd-DTPA) showing prominent perifocal edema (B), and a ring-enhanced lesion in the right cerebellar hemisphere below the tentorium (C, D).

guidance on December 10, 1990. Brownish-green pus was aspirated. Culture of the fluid demonstrated streptococci. His symptom improved soon after the operation with normalization of CT findings (Fig. 6). A two-staged operation on the PAVF was performed on February 4 and 25, 1991. He had a good postoperative course and no recurrence of the abscess has since been observed.

Discussion

HHT associated with PAVF was an underrecognized cause of brain abscess,⁷⁾ but several recent reports^{4,7,12)} suggest that these concurrences are relatively common.¹⁰⁾ However, brain abscesses complicated by HHT are usually supratentorial. Only one previous case of cerebellar abscess secondary to HHT has been reported.¹⁹⁾ Most (48–93%) of cerebellar abscesses are secondary to adjacent otic infections.^{1,14,16,20,22)} Our case of cerebellar abscess complicated with PAVF and HHT is extremely unusual,

Fig. 5 Left vertebral angiograms (A: anteroposterior view of arterial phase, B: lateral view of arterial phase, C: anteroposterior view of venous phase, D: lateral view of venous phase) showing an avascular area, but no vascular abnormality.

but the reason for the formation of the abscess in the posterior fossa is still unknown.

The first choice for treatment of brain abscesses is administration of antibiotics followed up by serial CT scans. Broad spectrum antibiotics should be used until specific culture data are available. However, the typical cerebellar abscess is likely to induce hydrocephalus, so must be regarded in a different category from supratentorial abscess. High mortality continues to be associated with cerebellar abscesses despite modern neuroimaging improvements,^{3,17,22)} which reflects the unpredictable and dangerous nature of this condition.

Various surgical techniques are used to brain abscesses including simple aspiration, external drainage, and/or total excision. Simple aspiration may result in a good outcome, but recurrence of the abscess may occur. Gruskiewicz *et al.*⁹⁾ recommended total excision for abscesses in the posterior fossa because the abscess may sometimes communicate through the dura with the focus of infection in the mastoid or pyramid. Metastatic cerebellar abscesses

Fig. 6 Postoperative CT scans (left: precontrast, 7 days later; right: postcontrast, 7 wks later) showing the size of the abscess had decreased considerably after the operation, and finally diminished.

like the present case are good candidates for aspiration. Ultrasound guidance was very effective to demonstrate the extent of the abscess and the position of the catheter.

PAVF should be eradicated because brain abscesses associated with untreated PAVF recurred until the PAVF was resected.^{4,8,18} Van Dellen *et al.*²² insisted that surgical drainage of the abscess alone is insufficient, and complete eradication of the source of sepsis and adequate administration of antibiotics is essential. The PAVFs in our patient were resected in two-staged operation because the PAVFs were bilaterally located. Recently, PAVF has been treated by endovascular embolization, which is suitable for multiple lesions as well as high risk patients.^{6,21}

Cerebellar abscess can occur secondary to HHT and PAVF. Complete eradication of the PAVF is essential because the brain abscess will sometimes recur unless this is achieved.

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Address reprint requests to: M. Ogino, M.D., Department of Neurosurgery, Saiseikai Utsunomiya Hospital, 911-1 Takebayashi-cho, Utsunomiya, Tochigi 321, Japan.