J Korean Med Sci 2002; 17: 407-11
ISSN 1011-8934

The Multiple Brain Abscesses Associated with Congenital Pulmonary Arteriovenous Malformations
: A Case Report

In this report, we descrbed a case of multiple brain abscesses associated with diffuse congenital pulmonary arteriovenous malformations (PAVM). Although the cases of brain abscesses associated with congenital PAVM are very rare, the brain abscess could be an initial clinical manifestation in asymptomatic PAVM as in the case presented in this report. PAVM may contribute to the development of a brain abscess by allowing easy bacterial access to systemic circulation through the right-to-left pulmonary vascular shunt, bypassing the filtering effect of the pulmonary capillaries. Hence, this association should be considered in cases with brain abscesses of undetermined etiologic factors.

Key Words : *Arteriovenous Malformations; Brain Abscess; Pulmonary Arteriovenous Malformations, Congenital*

Seok Han, Dong-Jun Lim, Yong-Gu Chung, Tai-Hyoung Cho, Seong-Jun Lim, Woo-Jae Kim, Jung-Yul Park, Jung-Keun Suh

Department of Neurosurgery, Medical College, Korea University, Seoul, Korea

Received : 31 May 2001
Accepted : 4 July 2001

Address for correspondence
Dong-Jun Lim, M.D.
Department of Neurosurgery, Korea University, Ansan Hospital, 516 Gojan-dong, Ansan 425-020, Korea
Tel : +82.31-412-5053, Fax : +82.31-412-5054
E-mail : djlim@kg21.net

INTRODUCTION

The well-known etiologic factors of brain abscess are otitis media, paranasal sinusitis, previous history of craniotomy or open head injury, and cyanotic congenital heart disease. Among these, cyanotic congenital heart disease is the most frequent underlying disease in pediatric brain abscesses (1, 2). More than 500 cases of pulmonary arteriovenous malformation (PAVM) have been reported as a rare cause of right-to-left shunt (3). Brain abscess could be precipitated by ill-fated embolization, finding a way around the filtering effect of pulmonary capillaries. The incidence of PAVM is rare when compared with that of cyanotic congenital heart disease, but the incidence of development of neurological complications are higher in patients with PAVM. Until now, more than 60 cases of brain abscess associated with PAVM have been reported.

This report describes a case of multiple brain abscesses associated with congenital PAVM with the literature review.

CASE REPORT

An 11-yr-old boy was presented with chief complaints of headache and nausea for a week. He had neither any remarkable past medical history nor family history. The initial neurological examination revealed that he was mentally drowsy and had right-side facial weakness. On admission, he was afebrile. His general physical examination did not present cyanosis, tachypnea, multiple small lesions on the mucosal surface of the mouth, or digital clubbing of the hands and feet. Laboratory evaluation was notable for a white blood cell count of 12,000/μL with normal differential counts, hemoglobin 16.7 g/dL, and erythrocyte sedimentation rate 7 mm/hr. Arterial blood gases were PaO_2 52.2 mmHg, $PaCO_2$ 26.5 mmHg, and HCO_3^- 19.5 mmol/L. Hypoxemia without $PaCO_2$ elevation was noted. An elevated level of calculated (A-a) DO_2 was also identified. The presence of shunt was suspected secondary to correction of hypoxia, which was not improved with supplementary oxygen. Nevertheless, an intra-cardiac shunt was not evident on transthoracic Doppler examination. Plain chest radiographs showed subtle vascular opacities in the right hilar region and no other abnormal densities in either lung fields (Fig. 1). The contrast-media-enhanced brain CT study demonstrated a 3.2 cm-sized, multi-lobulated, cystic septated mass in the left fronto-temporal region and another smaller cystic mass in the ipsilateral occipitoparietal area with a homogenous regular rim enhancement. The prominent low densities around the mass lesions were shown along the white matter tract, which induced midline shifting and contralateral cisternal obliteration (Fig. 2). He experienced generalized tonic seizures that lasted about 3 min while he was under medical control for his elevated intracranial pressure at the intensive care unit.

After the episode, his neurological conditions deteriorated into a mentally stuporous state due to tentorial herniation. Therefore, an emergency craniotomy for the removal of the larger cystic mass was undertaken. After the dura had been opened, the abscess cavity was exposed via the trans-cortical approach. The general parenchymal swelling was subsided after the sufficient drainage of yellowish pus with a foul odor. The abscess cavity showed premature capsular formations and its culture revealed the growth of Moraxella species. Hypoxia sustained throughout the operation, but positive end expiratory pressure (PEEP) was not applied because of the possibility of pulmonary vascular shunt. The patient was managed with intravenous administrations of ceftriaxone and vancomycin, to which the pathogen was sensitive, and mannitol for the control of increased intracranial pressure. His neurologic conditions improved with the management. On the post-operative fifth day, he was mentally alert without hemiparesis. After recovery, the contrast-media-enhanced chest CT study and pulmonary angiography confirmed the presence of diffuse bilateral PAVM, most prominently in the right lower lobe (Fig. 3, 4). Postoperatively, brain CT with contrast-media enhancement was obtained on a weekly basis. Abscess aspiration was performed 3 weeks after the operation, because of enlargement of temporal and occipitoparietal cavities on a follow-up scan, although this was not associated with any clinical deterioration. Antibiotics had been used for 12 weeks. He made a satisfactory recovery and serial scans confirmed gradual resolution of abscess cavities (Fig. 5). A few treatment options for the diffuse PAVMs are available for now. We think that transcatheter embolotherapy may be helpful to reduce the risk of neurologic complications, so he has now been referred for that.

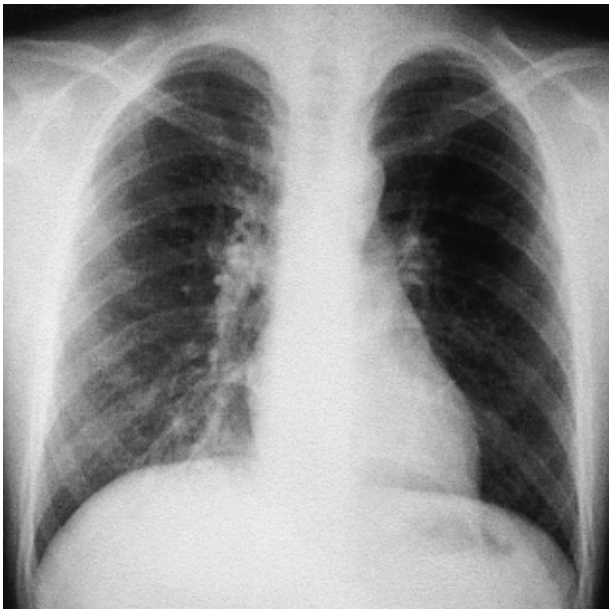

Fig. 1. The plain chest anteroposterior view shows only subtle vascular opacities in the right hilar region.

DISCUSSION

The incidence of brain abscess in children was reported to be 4% of the total number of patients with a space-occupying intracranial lesion (1). Cyanotic congenital heart disease

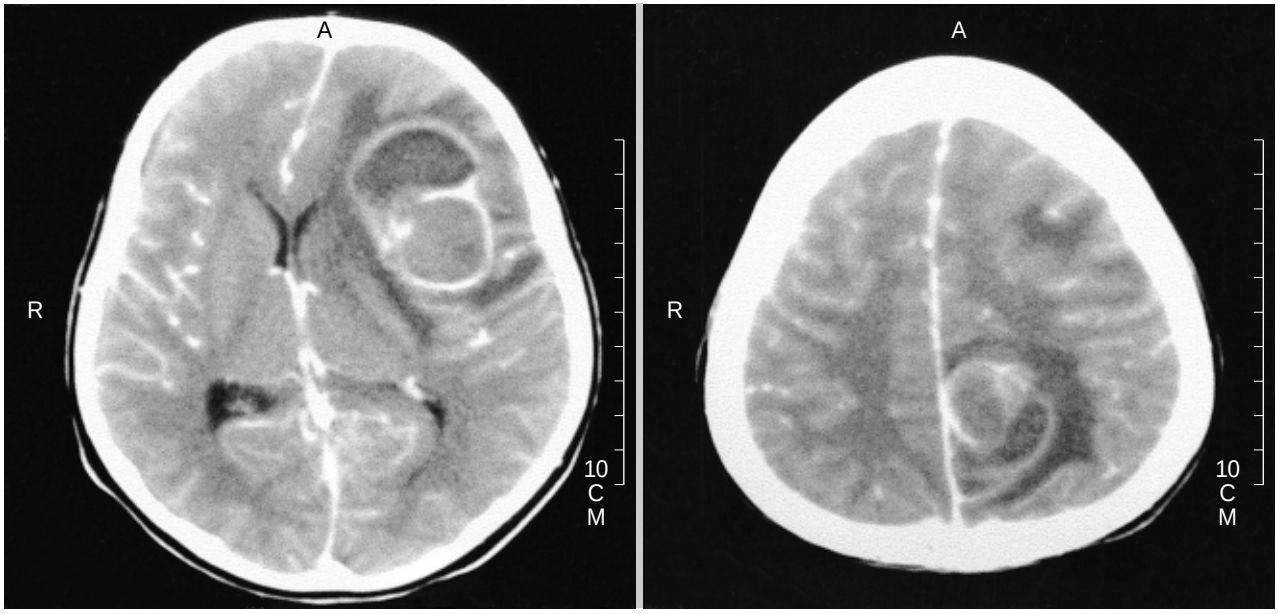

Fig. 2. The brain CT with contrast enhancement demonstrates a multilobulated, septated cystic mass in the left fronto-temporal region and another smaller cystic mass in the ipsilateral occipital area with a regular homogenous rim enhancement.

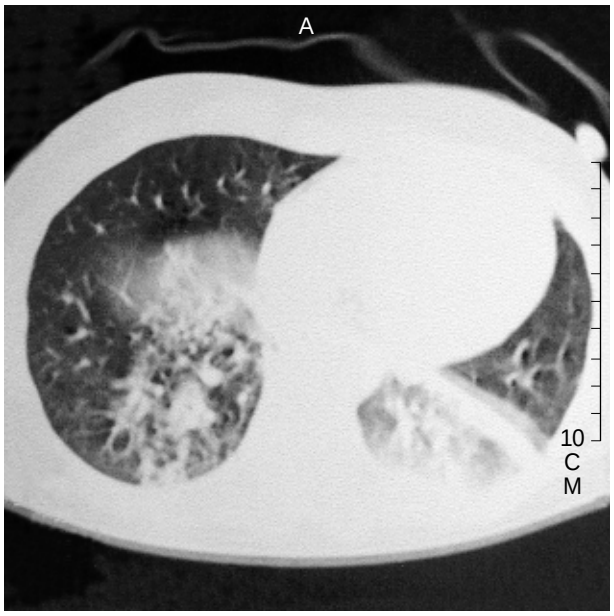

Fig. 3. The chest CT with contrast enhancement reveals prominent vascular opacities in both hilar regions.

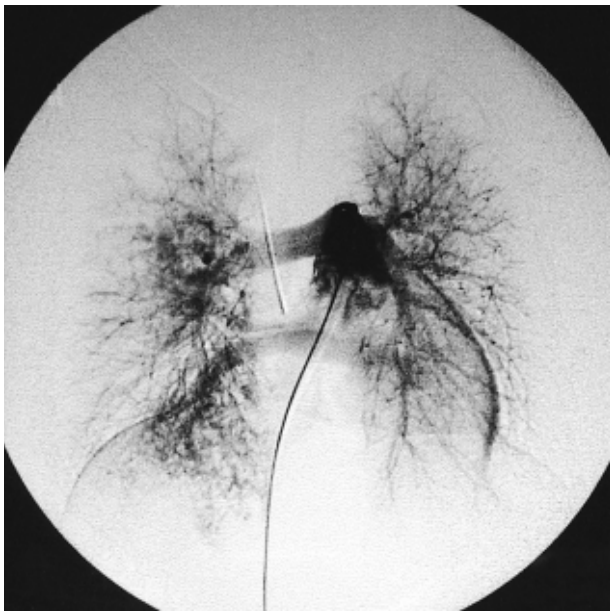

Fig. 4. As compared with vascularity of the right upper lobe, many capillaries are visible in the right lower lobe in the early arterial phase. In the mid-arterial phase, the diffuse scattered arteriovenous malformations are demonstrated and pulmonary vein and filling of the left atrium are also visible by the shunt.

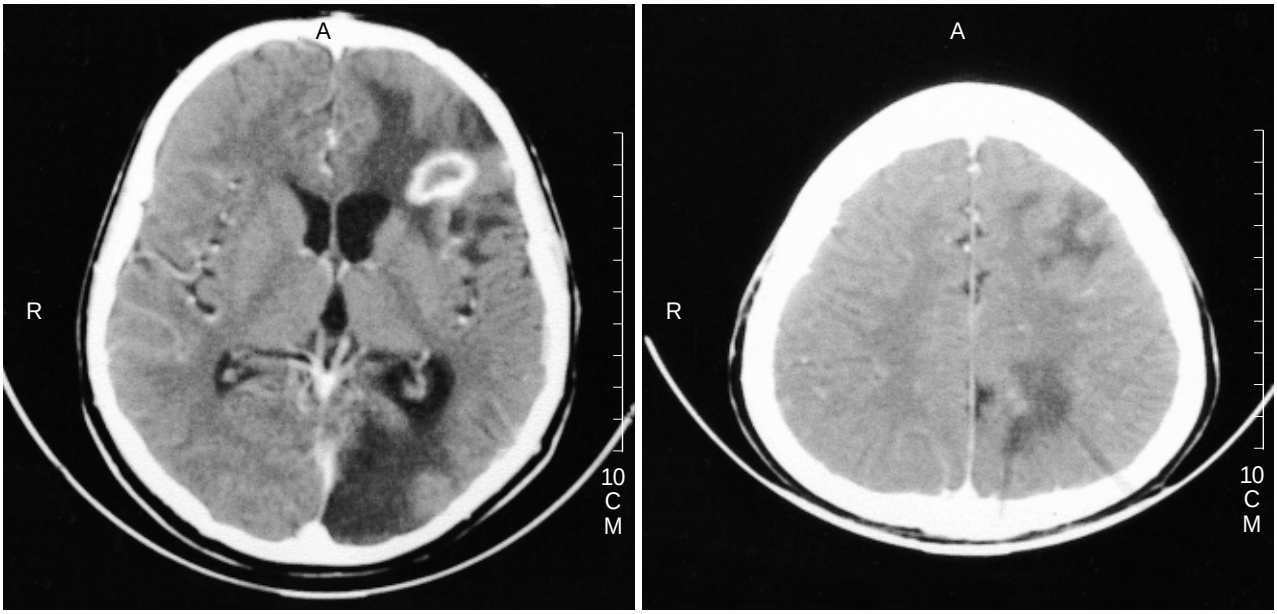

Fig. 5. The brain CT with contrast enhancement at eleven weeks following operation shows prominent reduction of abscess cavities.

is known to be the most frequent predisposing factor of brain abscess in children, taking up 30-50% of all factors (1, 2). Churton (5) first described PAVM in a 12-yr-old boy and Reading (6) first reported a case of brain abscess with signs of the classic triad (cyanosis, polycythemia, and clubbing of the finger and toes) and with an association with PAVM revealed after a postmortem examination in 1932. The vast majority of PAVM is of congenital (3), and currently-accepted theory of its origin is incomplete degeneration in the septa that divide the primitive connection between the arterial and venous plexuses at the level of pulmonary buds (6). Therefore, it is characterized by a pathological con-

nection between an afferent artery and one or more efferent veins without an interposed capillary bed (2). Congenital PAVM may occur singularly, in multiples, or bilaterally. Thirty-six percent of patients with a single lesion and 57% of those with multiple lesions are associated with Rendu-Osler-Weber syndrome (hereditary hemorrhagic telangiectasia) and conversely, 15% of the patients with Rendu-Osler-Weber syndrome have PAVM (3, 4). More than 90% of patients with Rendu-Osler-Weber syndrome who developed brain abscess or meningitis, however, were reported to be associated with PAVM (4, 7). The case in this report had no family history nor clinical findings that correspond to Rendu-Osler-Weber syndrome-recurrent epistaxis, gastrointestinal bleeding, and mucocutaneous telangiectasis. If the amount of right-to-left shunt is more than 25% of the total systemic circulation, it may lead to clinical manifestations of PAVM such as dyspnea, cyanosis, chronic fatigue, and hemoptysis (8). Asymptomatic PAVM is common in cases with a small amount passing through the right-to-left shunt (9, 10). Neurological complications develop in 8 to 41% of patients with PAVM, which include hypoxic encephalopathy, paradoxical embolism, transient ischemic attack, intracerebral hemorrhage, and brain abscess (4, 9-11). Hence, a brain abscess in asymptomatic PAVM could be clinically manifested at the initial stage, as evidenced by this case. Of the neurological complications of PAVM, a brain abscess occurs in 5 to 10% of patients with PAVM and the expected risk of development of a brain abscess in patients with PAVM is approximately 1,000 times greater than that of the CNS infection occurring in the general population (4). Septic embolization circulating in the pulmonary capillaries through the right-to-left shunt, spreading into the areas of cerebral microinfarction may explain the mechanism responsible for the development of brain abscess in patients with PAVM. Momma, et al. (7) reported that the supratentorial region, especially parietal lobe, is the most common site of abscess, reflecting the aspect of septic embolization in the blood circulation, and 7% of the multiple abscess cases with PAVM occurs in this region. Additionally, brain abscesses associated with hematogenous dissemination arising from a distant site are often deep-seated, and relatively poorly encapsulated on initial diagnosis. Consequently, these have unfavorable prognosis (1). Mamelak et al. (12) advocated the principles in the management of multiple brain abscesses: 1) if multiple ring-enhancing lesions are found, emergency surgery for all abscesses greater than 2.5 cm in diameter or causing a marked mass effect should be undertaken, either by excision or preferably by stereotactic aspiration; 2) broad-spectrum antibiotics should be utilized until culture results are available, and then antibiotics can be tailored adequately to these results. Afterward, antibiotics should be continued for a minimum of 6-8 weeks or often for over a year in immuno-compromised patients. Repeated surgical drainage should be undertaken if there is an enlargement of an abscess after 2 weeks of treatment or when clinical deterioration occurs, or if an abscess fails to diminish in size after 4 weeks of treatment with antibiotics. The application of this combined approach should yield cure rates of higher than 90% in patients with multiple brain abscesses, a result similar to that expected in patients with solitary lesions (1).

Worsening hypoxia during surgery on patients with PAVM appears to be very rare. PEEP, which normally keeps small bronchioles and alveoli open, also compresses the alveolar capillaries, reducing the circulation to larger vessels and the shunt through PAVM will worsen. Thus, if there is a pulmonary vascular shunt, PEEP should be discontinued, even if this results in a subnormal PaO_2. In diagnosing PAVM, arterial blood gas analysis is helpful and 95% of patients will have an abnormal finding on the plain chest radiograph (3, 4, 9). Chest CT with contrast-media enhancement study and pulmonary angiography will confirm the diagnosis. The symptoms arising from PAVM progress in 26% of these cases and the mortality rate from these complications reaches 11%. At any rate, all cases should be considered for treatment (3, 9). Surgical resection or lung transplantation is of limited value in patients with multiple lesions as balloon and coil embolization has become an acceptable method of management (11, 13).

This report described a case of multiple brain abscesses associated with congenital PAVM. PAVM should be considered as an etiologic factor in cases of chronic hypoxemia combined with appearance of brain abscess of unknown etiology.

REFERENCES

1. Ciurea AV, Stoica F, Vasilescu G, Nuteanu L. *Neurosurgical management of brain abscesses in children. Child's Nerv Syst 1999; 15: 309-17.*
2. Wong TT, Lee LS, Wang HS, Shen EY, Jaw WC, Chiang CH, Chi CS, Hung KL, Liou WY, Shen YZ. *Brain abscesses in children - a cooperative study of 83 cases. Child's Nerv Syst 1989; 5: 19-24.*
3. Walder LA, Anastasia LF, Spodick DH. *Pulmonary arteriovenous malformations with brain abscess. Am Heart J 1994; 127: 227-32.*
4. Hall WA. *Hereditary hemorrhagic telangiectasia (Rendu-Osler-Weber disease) presenting with polymicrobial brain abscess. J Neurosurg 1994; 81: 294-6.*
5. Churton T. *Multiple aneurysms of pulmonary artery. BMJ 1897; 1: 1223.*
6. Reading B. *Case of congenital telangiectasia of the lung, complicated by brain abscess. Tex State J Med 1932; 28: 462.*
7. Momma F, Ohara S, Ohyama T, Moto A, Okada H, Harada H. *Brain abscess associated with congenital pulmonary arteriovenous fistula. Surg Neurol 1990; 34: 439-41.*
8. Chang UG, Kim DG, Yang HJ, Lee HK, Choi KS, Han DH. *Brain abscess associated with pulmonary arteriovenous malformation. J Korean Neurosurg Soc 1992; 21: 895-8.*

9. Brydon HL, Akinwunmi J, Selway R, UI-Haq I. *Brain abscesses associated with pulmonary arteriovenous malformations. Br J Neurosurg 1999; 13: 265-9.*
10. Caroli M, Arienta C, Rampini PM, Balbi S. *Recurrence of brain abscess associated with asymptomatic arteriovenous malformation of the lung. Neurochirurgia 1992; 35: 167-70.*
11. Faughnan ME, Lui YW, Wirth JA, Pugash RA, Redelmeier DA, Hyland RH, White RI. *Diffuse pulmonary arteriovenous malformations : characteristics and prognosis. Chest 2000; 117: 31-8.*
12. Mamelak AN, Rosenblum ML, Obana WG, Mampalam TJ. *Improved management of multiple brain abscess: a combined surgical and medical approach. Neurosurgery 1995; 36: 76-85.*
13. Suresh CG, Coupe MO, Jegarajah S. *Recurrent cerebral abscesses 20 years before recognition of multiple pulmonary arteriovenous malformations. Br J Clin Pract 1995; 49: 105-6.*