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Case Report

A Multi-organ Abscesses Including Brain Caused by a Congenital Pulmonary Arteriovenous Fistula

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In cases of brain or other organ abscess, the causative etiology or disease are not always definable. We report a case of brain, renal, and possibly lung abscesses in a middle aged woman. After close, stepwise surveillance of possible etiologic factors, we covered out a small solitary pulmonary arteriovenous fistula without any pulmonary symptoms and successfully occluded the fistula via endovascular approach. The congenital pulmonary arteriovenous fistula should be bear in mind as a cause of repeated, multiple systemic infective source spray and be pursued despite of negative initial baseline studies.

KEY WORDS : Abscess · Arteriovenous fistula · Brain · Congenital · Kidney · Lung.

Introduction

The causes of brain abscess are numerous. In practical field, the contagious propagation and distant septic foci inoculation are two important causative factors. But in many cases, physicians are failed to, or ignored to verify the causative factors and they make the abscess itself as the only target disease. For example, the sinusitis, one of the major cause of brain abscess, may be caused by hereditary disorder such as primary mucociliary transport failure[16]. The pulmonary arteriovenous fistula has abnormal shunting of venous blood into systemic circulation and is well known septic original source[3,4,5,9,10,12]. In case of large, definite pulmonary arteriovenous fistula, simple chest film can reveal its abnormality and it can be ruled out without difficulty. We report a rare case of simu-ltaneous multiple manifestation of multi-organ abscesses including brain, kidney and possibly lungs. After careful and close surveillance of possible causes of septic emboli, we found small solitary arteriovenous fistula and definitely treated using endovascular approach.

Case Report

Symptom summary and initial workup with tentative diagnosis

A 41-year-old woman presented with 3-week history of headache with vomiting and 8-month history of dull right flank pain. Medical history revealed neither chronic con-sumptive disease, such as diabetes, hepatitis, tuberculosis nor immunocompromised condition. During last 1 year, her body weight loss was about 10kg. Her initial vital signs were stable without fever. Except for knocking tenderness at right costovertebral angle, other physical examination showed no abnormality. Her skin contour was normal. She was not cyanotic or tachypneic. Neurologically her mental status was alert and other focal neurologic deficits were not detectable. She complained dull headache only. Laboratory surveillance showed some following abnormalities. The hemoglobin was 7.3g/dL, the white blood cell count was 19,710/ul with segment form predominance(91.1%), the platelet count was 754,000/ul. ESR was moderately increased with the value of 60mm/hr. CRP was increased also as 7.63mg/dL. Bactriuria with 10~29WBC/HPF and hematuria 10~29RBC/HPF were detected. Direct and indirect Coomb's test were reported positive and negative, respectively. Routine blood chemistry and arterial blood gas analysis showed no abnormality. The contrast enhanced abdomen CT study demonstrated diffusely enlarged right kidney with multilobulated honeycomb shaped eccentric mass lesion. The normal calyx structure was markedly distorted(Fig. 1A). Another cut showed additional cystic mass. Considering above laboratory (anemia, hematuria), physical (weight loss) and radiologic (aggressively destructive renal mass) all led to tentative diagnosis of renal cell carcinoma. Brain CT and MRI showed cystic lobulated mass with irregularly thickened wall and geographic surrounding edema was noted on left occipital lobe(Fig. 1B). Considering abdominal mass lesion, we suspected this brain lesion to be a renal origin metastatic tumor.

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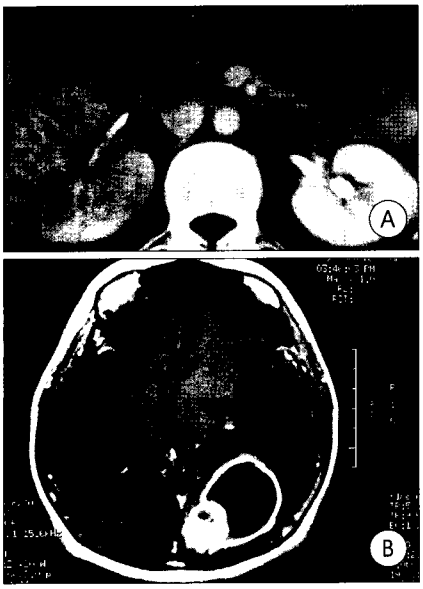

Fig. 1. A : An abdomen computed tomography shows a marked enlarged right kidney with multilobulated septation and destructed normal calyx pattern. B : The brain magnetic resonance image with enhancement shows a well enhancing cystic tumor mass having a irregularly thickened wall on left occipital lobe.

Operation and medical treatment courses

For more symptomatic brain lesion, craniotomy was done firstly. Intraoperatively, foul odoured thick abscess was noted. The wall of abscess was removed and abscess was removed, irrigated and drained in usual manner. Gram stain and further bacteriological culture studies were inert. At that time, we still convinced that the renal cell carcinoma induced immunologic decline led to brain abscess formation. We deferred curative surgery of renal cell carcinoma until completion of full 6 week antibiotic therapy. After combined antibiotic therapy, the brain abscess was successfully collapsed(Fig. 2A). Follow up abdominal CT scan was checked for immediate preo-perative check up of renal tumor. Interestingly, previous huge renal mass was near completely healed and the normal renal calyx contour was restored(Fig. 2B). Retrogradely thinking, the renal mass was not a kind of malignancy but abscess induced pseudotumor, which was effectively eradicated by combined antibiotic therapy for brain abscess.

Surveillance for hidden causative factors and curative therapy

Her general and neurologic condition were much improved after surgery and antibiotic therapy. The laboratory abnormalities including inflammation markers were corrected normally. But considering her age and previously healthy condition, the simultaneous multifocal abscess formation was very rare event. So we decided to seek another possible causative factors. Firstly, several laboratory tests were performed to rule out the possibility of immunologic disease (HIV ELISA, CD4, CD8, immunoglobin assay, complements assay, CH_{50}) or autoimmune disease (anti-DNA antibody, anti-Sm, antinuclear antibody). All available data showed no definite abnormalities. After close surveillance of various causes of septic foci reproduction, we suspected pulmonary arteriovenous fistula(PAVF) to be the most possible cause. We reviewed the simple chest film, but could not find any evidence of abnormality. Because in the initial radiological report, the lesions were diagnosed as "multiple healed tuberculosis", we informed the details of the patient's clinical history and asked reevaluation of possibility of PAVF. The chest radiologist found that the enhancement of small nodules, which were connected to serpiginous linear opacities that were suspicious artery and adjacent small venous structure with short interconnection, highly suggesting PAVF(Fig. 3A). More detailed examination revealed another small enhancing nodularities on peripheral lung field suggesting multiple septic emboli, rather than healed tuberculosis. For rule out rare hereditary disease accompanying with PAVF, hereditary hemorrhagic telangiectasia (Rendu-Osler-Weber disease), physical examination (various mucosal telangiectasias) and detailed history taking (epistaxis, family history of telangiectasia) were performed. The results were all negative. For confirmation of small solitary PAVF, we checked pulmonary angiography and finally disclosed 1.8×1.5cm sized simple type AVF on right lower lobe of lung(Fig. 3B). Via endovascular approach, the fistula site was successfully occluded using Hilal platinum embolization microcoil(Cook Inc., Bloomington, IN, USA)(Fig. 3C). During 1.5year follow up, one brain CT and two chest CT showed stable brain condition and well positioned coil without new fistular dilatation, respectively.

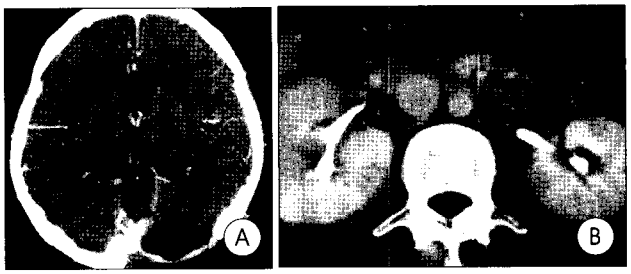

Fig. 2. A : The postoperative brain computed tomography shows a successfully removed and collapsed previous abscess cavity. B : After 6 weeks of antibiotic therapy for eradication of the brain abscess, the abdomen computed tomography reveals normalization of renal calyx pattern and much reduction of multilobulated cystic lesion. This looks like a simultaneous infectious abscess cavity, rather than a tumor cyst.

Discussion

Pulmonary arteriovenous fistula(PAVF)

The PAVF is a kind of congenital anomaly and is consi-

Fig. 3. A : The contrast enhanced chest computed tomography shows a simultaneous enhancement of a small, suspicious artery and surrounding small venous structure with short interconnection, suggesting pulmonary arteriovenous fistula (PAVF) (arrow). B : The selective pulmonary angiography reveals simultaneous visualization of both pulmonary artery and vein through PAVF connecting nodule, adjacent to the bronchovascular bundles at lower lobe of right lung (arrow). C : After occlusion of PAVF nodule using a Hilal coil (arrow), an abnormal fistulous shunting of blood was completely disappears.

dered to be caused by incomplete degeneration in the septa that divide the primitive connection between the arterial and venous plexuses at the level of pulmonary buds. This defect in the terminal capillary loops results in dilatation and formation of thin-walled vascular sacs. Although it is assumed that the vascular defects are present at birth, they are seldom manifested clinically until adult life when the vessels have been subjected to pressure over several decades[3]. More than 70% of PAVFs are simple and have a single feeding artery and a single draining vein. Complex form of PAVFs, which have numerous feeding arteries and multiple draining veins, account for 20% of all cases. In less than 10% of cases, a diffuse pattern may be present[7,9]. It has similar gender predominancy and its symptomatic manifestations are not recognized until the third or fourth decade of life. Most patients with PAVF present with a round or lobulated subpleural mass, with 70% of the masses located in the lower lobes[1]. Approximately 30~50% of cases of patients with PAVFs have multiple lesions. In PAVF, special consideration should be given to hereditary hemorrhagic telangiectasia(HHT) or Rendu-Osler-Weber disease. The HHT is an autosomal dominant disorder characterized by recurrent episode of epistaxis and gastrointestinal hemorrhage[5]. In about 20~30% cases of PAVF, it can be accompanied with HHT. The pathophysiologic fundamental is right-to-left shunting of blood and it results in hypoxemia, paradoxical emboli and pulmonary hyper-tension. In large sized PAVF, sh-unting induced hypoxemia symp-toms such as dyspnea, cyanosis, fatigue or pulmonary hypertension elicited hemoptysis are more frequently encountered. But in case of small one, paradoxical emboli induced remote manife-station is more predominant. Chest CT scan with contrast enhancement and MRI imaging are adv-ocated for screening study, but confirmative diagnosis and treatment planning are purely depend upon selective pulmonary angiography[15].

Poor results of conservative management with approximately 30% of mortality or morbidity urge to treat aggressively[9]. Embolization with coils or detachable balloon can achieve occlusion of shunted site less invasively and it gradually replaces traditional thoracotomy based resection treatment[11,14]. Frequent follow up of treated patient is necessary because PAVFs tend to increase both in number and in size over time[13]. According to recent review, the success rate of embolization therapy is over 98% and embolic device migration and/or recanalization rate is about 2~4%[8].

Neurologic complication of PAVF

The neurologic manifestation of PAVF is not uncommon. According to Mayo clinic experience, it reported up to 34% incidence[13]. Various neurologic symptoms are explained by two basic pathophysiologic mechanisms. The first one is polycythemia induced sludging of blood flow and chronic hypoxia induced infarction. They eventually lead to hypoxic encephalopathy, transient ischemic attack, hemiparesis, seizure or hemorrhagic conversion. The second additional one is paradoxical emboli induced abscess and it more commonly encountered. The systemic venous blood are continuously "shunted" via PAVF and resultantly bypass macrophages govern purifying system. Now, the septic sources in this venous blood can reach any site of body, can lodge microinfarction area and finally start abscess formation. Among various areas of the body, the brain is one of the system with poor macrophage(microglia) governed scavenge system. So brain abscess is common manifestation[2,4,6,9,10,12].

The expected risk for developing brain abscess in patients with HHT is approximately 1,000times greater than the risk of for developing CNS infection in the general population[2,10].

As shown by this case and other reports, PAVF can often by asymptomatic with brain abscess being the first manifestation[4-6,12]. Previous report stressed that the smaller size of PAVF does not preclude septic embolization to the brain[5] and this emphasis clearly reconfirmed by our case experience with multi-organ involvement including brain.

Importance of diagnostic suspicion

In this case, we present our diagnostic steps in detail manner. The multiple abscess without any immunologic deficiency evidence led us stepwise diagnostic workup. Despite of her normal chest PA film and lack of any

pulmonary symptoms, we checked enhanced chest CT for rule out PAVF. Initially, without any clinical information, the radiologist misdiagnosed the lesion as a healed tuberculosis. However, providing a detailed, careful information about clinical findings and history to the radiologist, we could confirm a valuable diagnosis and eventually cured the original chest lesion, PAVF. We stress again the importance of diagnostic suspicion and interdepartmental communication.

Conclusion

We successfully treated pulmonary arteriovenous fistula(PAVF) induced multi-organ (brain, kidney and possibly lungs) abscess with neurosurgical craniotomy, appropriate antibiotics and endovascular occlusion therapy. In cases of multi organ abscess without known causative factors, the PAVF should be suspected, aggressively studied and definitely treated.

References

1. Dines DE, Arms RA, Bernatz PE, Gomes MR : Pulmonary arteriovenous fistulas. **Mayo Clin Proc 49** : 460-465, 1974
2. Dyer NH : Cerebral abscess in hereditary haemorrhagic telangiectasia : report of two cases in a family. **J Neurol Neurosurg Psychiatry 30** : 563-567, 1967
3. Fraser RS, Pare JAP, Fraser RG, Pare PD : **Synopsis of diseases of the chest**, ed 2. Philadelphia : W. B. Saunders, 1994, pp277-279
4. Fuentes Pradera MA, Otero Candelera R, Ortega Ruiz F, Franco E : Cerebral abscess as first manifestation of a familial pulmonary arteriovenous fistula. **Arch Bronconeumol 35** : 407-409, 1999
5. Hall WA : Hereditary hemorrhagic telangiectasia (Rendu-Osler-Weber disease) presenting with polymicrobial brain abscess. Case report. **J Neurosurg 81** : 294-296, 1994
6. Han S, Lim D, Chung Y, Cho T, Lim S, Kim W, et al : The multiple brain abscessed associated with congenital pulmonary arteriovenous malformations : A case report. **J Korean Med Sci 17** : 407-411, 2002
7. Higgins CB, Wexler L : Clinical and angiographic features of pulmonary arteriovenous fistulas in children. **Radiology 119** : 171-175, 1976
8. Khurshid I, Downie GH : Pulmonary arteriovenous malformation. **Postgrad Med J 78** : 191-197, 2002
9. Natarajan K, Khabiri H, Jung S : Case 3. Pulmonary arteriovenous malformation (PAVM) : paradoxical embolism through the arteriovenous fistula can cause brain abscess and infarct. **AJR Am J Roentgenol 175** : 857-858, 2000
10. Press OW, Ramsey PG : Central nervous system infections associated with hereditary hemorrhagic telangiectasia. **Am J Med 77** : 86-92, 1984
11. Remy J, Remy-Jardin M, Giraud F, Wattinne L : Angioarchitecture of pulmonary arteriovenous malformations: clinical utility of three-dimensional helical CT. **Radiology 191** : 657-664, 1994
12. Shibasaki I, Kase K, Ohtsuka A, Nakayama M : A case of pulmonary arteriovenous fistula associated with cerebellar abscess. **Kyobu Geka 52** : 512-515, 1999
13. Swanson KL, Prakash UB, Stanson AW : Pulmonary arteriovenous fistulas : Mayo Clinic experience, 1982-1997. **Mayo Clin Proc 74** : 671-680, 1999
14. Taylor BG, Cockerill EM, Manfredi F, Klatte EC : Therapeutic embolization of the pulmonary artery in pulmonary arteriovenous fistula. **Am J Med 64** : 360-365, 1978
15. White RI Jr : Pulmonary arteriovenous malformations : how do we diagnose them and why is it important to do so? **Radiology 182** : 633-635, 1992
16. Yoon SH, Lee HJ, Yi JS, Yang JH, Lee IW, Song KS, et al : A case of brain abscess in a patient with primary mucociliary transport failure-A case report. **J Korean Neurosurg Soc 30** : 1430-1434, 2001