A 50-Year-Old Man With Clubbing Has Seizures After a Dental Procedure*

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A 50-year-old white man presented to the emergency department with right-sided focal seizures with secondary generalization. The first episode was 3 h prior to hospital admission; his daughter, who called an ambulance, witnessed the second episode. The patient worked as an engineer and denied any history of smoking or alcohol abuse. His medical history was unremarkable, with specifically no known convulsive or respiratory disorders. He was not taking any medications. He had no previous surgical history other than an uneventful root canal procedure 4 weeks prior. He was known to have clubbing, and abnormal chest radiograph findings since childhood, which had never been investigated. On further questioning, he reported recurrent spontaneous epistaxis and a long history of shortness of breath and chest pain with exertion, although he remained active.

Physical Examination

On admission, the patient appeared well, with no respiratory distress and was neurologically intact. He looked plethoric and had marked clubbing of all fingers (Fig 1) and toes. Vital signs were as follows: body temperature, 37.8°C; pulse rate, 96 beats/min; BP, 118/68 mm Hg; respiratory rate, 28 breaths/min; and pulse oximetry on room air at rest, 90%. The rest of the physical examination was unremarkable, with specifically no bruit over the chest and no evidence of mucocutaneous telangiectasias.

Laboratory Data and Radiographic Findings

CBC count revealed marked polycythemia (hemoglobin, 21.6 g/L; HT 0.653 L/L) without leukocytosis. Blood gas values while the patient breathed room air showed hypoxemia with respiratory alkalosis (pH 7.51; Pco2, 29 mm Hg; Po2, 59 mm Hg; bicarbonate, 24 mmol/L). ECG revealed sinus tachycardia with right-axis deviation.

Cerebral CT revealed an irregular ring-enhancing lesion involving the left parietal lobe. Chest radiography was performed (Fig 2) followed by unenhanced helical thoracic CT (Fig 3).

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Figure 1. Marked clubbing of the fingers.
What is the most likely diagnosis?

Suggest diagnostic modalities and therapeutic intervention.
Diagnosis: Pulmonary arteriovenous malformation complicated by cerebral abscess after a dental procedure.

Diagnostic modalities and therapeutic intervention: Shunt fraction measurement, contrast echocardiography, and pulmonary angiography with vascular embolization.

Imaging Studies

On chest radiography (posteroanterior and lateral), there is a large (9 cm) lobulated mass in the right lower lobe with several associated large vessels connecting with the right pulmonary hilum. No other abnormalities are seen. On CT scan of the thorax, a large multilobulated mass is present in the right lower lobe. There are multiple enlarged, tortuous feeding arteries entering the aneurysm, as well as a massive draining vein leading to the left atrium.

Discussion

First described in 1897, pulmonary arteriovenous malformation (PAVM), although uncommon, should be included in the differential diagnosis of pulmonary nodules, especially when associated with hypoxemia. PAVMs are direct communications between pulmonary arteries and veins, usually with an intervening thin-walled aneurysm. PAVMs may be present at birth, but in most cases they remain unrecognized and asymptomatic until the fourth to sixth decade of life. In approximately 80% of patients with PAVMs, hereditary hemorrhagic telangiectasia is the underlying disease, whereas most of the remainder are idiopathic. Due to the resulting right-to-left shunt, PAVMs may produce dyspnea, cyanosis, clubbing, and pulmonary bruit, although this classical presentation is uncommon. In patients with PAVMs, digital clubbing has been described only in those with large shunt and severe hypoxemia. The most common symptom is dyspnea on exertion, which occurs in approximately 50% of patients with PAVMs. Hemorrhagic complications, either massive hemothysis or spontaneous hemothorax, occur in approximately 15% of untreated patients, and can be fatal.

The association between PAVM and CNS complications has been well documented. These disabling and life-threatening conditions can be the first clinical manifestation of PAVMs, as in our case, and PAVMs are an increasingly recognized cause of stroke in younger patients without vascular risk factors. The prevalence of neurologic complications was best documented by White et al, who evaluated 76 consecutive PAVM patients and reported previous stroke in 18% of patients, cerebral abscess in 9%, seizures in 8%, transient ischemic attacks in 37%, and migraine headaches in 43%. The most likely mechanism for stroke is paradoxical embolization through direct artery-to-vein connections. The loss of the capillary filter is presumed to account for embolization of infected material as well, with development of cerebral abscess. Other predisposing factors for cerebral abscess in these patients might be hypoxemia, polycythemia, and previous cerebral infarction. Animal models have shown that intact brain parenchyma is relatively resistant to infection and there must be a preexisting area of hypoxia or ischemia to enable cerebral abscess formation. There is probably a correlation between risk for neurologic complications and feeding artery diameter. The prevalence of previous cerebral infarction appears to be greatest in patients with multiple and diffuse PAVMs compared to patients with a single PAVM. Cerebral infarction occurs almost exclusively in patients with feeding artery diameters > 3 mm, although it is unclear that there is a minimal feeding artery diameter required for development of cerebral abscess.

Although pulmonary angiography remains the “gold standard” for diagnosing PAVMs, the use of less invasive methods, such as shunt fraction measurement and contrast echocardiography, are useful for screening patients with suspected PAVMs. Agitated saline solution transthoracic contrast echocardiography is a sensitive test for the detection of intrapulmonary shunt, which is demonstrated by delayed appearance (vs early appearance in intracardiac shunt) of bubbles in the left atrium. Screening for PAVMs should be considered in patients with unexplained stroke, cerebral abscess, massive hemothysis, and spontaneous hemothorax. In addition, it is recommended to screen all patients with hereditary hemorrhagic telangiectasia for PAVMs, which are present in 15 to 33% of this population.

First performed in the late 1970s, transcatheter embolotherapy of PAVMs has become the treatment of choice, and has replaced traditional surgical management. Embolization is currently recommended for all PAVMs with a feeding artery of ≥ 3 mm to prevent stroke, cerebral abscess, massive hemothysis, and spontaneous hemothorax. The most common complication of transcatheter embolotherapy is self-limited pleuritic chest pain in approximately 20%, with major complications (air embolism and device paradoxical embolization) occurring in < 5%. In addition, the use of prophylactic antibiotics for dental work and other bacteremic procedures is recommended in all patients with PAVMs (even once embolized) for prevention of cerebral abscess. After successful embolization, patients should be followed up with oxygen shunt testing and unenhanced helical
CT 1 year after embolotherapy and then at least every 3 to 5 years thereafter, to assess for PAVM reperfusion and to look for the development or growth of other PAVMs.

The patient underwent stereotactic aspiration of the cerebral abscess. Gram stain of the aspirated fluid revealed many polymorphonuclear and positive cocci in pairs, and cultures subsequently grew fully sensitive *Streptococcus milleri*. On the seventh day in hospital, the patient underwent diagnostic pulmonary angiography (Fig 4, top), confirming a large, complex right lower lobe PAVM supplied by nine feeding arteries. Transcatheter embolotherapy was subsequently performed, with no residual aneurysmal opacification on completion angiography (Fig 4, bottom). There were no immediate complications. Two days after the procedure, right pleuritic chest pain and a pulmonary infiltrate with pleural effusion developed, suggestive of a postembolization infarct in the right lower lobe. He recovered completely over a 3-week period. The patient was discharged on the 14th day of hospital admission, receiving phenytoin and IV antibiotics.

One year after embolization, the patient was clinically well, with markedly improved exercise tolerance and no recurrent seizures. He had an oxygen shunt test done that showed no residual shunt. Unenhanced CT of the thorax showed involution of the aneurysmal sac (Fig 5).

**Clinical Pearls**

1. PAVMs can lead to CNS complications, such as cerebral abscess and strokes.
2. Screening for PAVMs is recommended in patients with unexplained stroke, cerebral abscess, massive hemothysis, spontaneous hemothorax, and all patients with hereditary hemorrhagic telangiectasia.

3. Noninvasive diagnostic modalities, such as shunt fraction measurement and contrast echocardiography, are useful for screening.

4. Transcatheter embolotherapy is the treatment of choice and recommended for all PAVMs with a feeding artery of ≥ 3 mm.

5. Use of prophylactic antibiotics for bacteremic procedures is recommended in all patients with PAVMs, even after successful embolotherapy.

6. Follow-up to ensure long-term success of embolotherapy and rule out new or growing PAVMs is essential. This should include oxygen shunt testing and unenhanced helical CT 1 year after embolotherapy and then at least every 3 to 5 years.

Selected Readings
Churton T. Multiple aneurysms of the pulmonary artery. BMJ 1897; 1:1223–1225


