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Brain abscess as the first clinical manifestation of multiple pulmonary arteriovenous malformations in a patient with hereditary hemorrhagic telangiectasia (Rendu-Osler-Weber disease)

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Abstract

In this report we described a case of a cerebral abscess that developed in presence of asymptomatic pulmonary arteriovenous malformations (PAVMs) in a 53-year-old woman with hereditary hemorrhagic telangiectasia (HHT). The brain abscess was aspirated with good clinical result and the arteriovenous fistulae qualified for transcatheter embolotherapy.

Each patient suspected to suffer from HHT should be diagnosed for the presence of visceral vascular malformations. Most of them are found in the lungs, liver and brain. Early diagnosis and treatment of PAVM prevent the occurrence of severe neurological complications such as brain stroke or brain abscess. Cases of a cerebral abscess in adults of unexplained etiology should raise the suspicion of an asymptomatic PAVM.

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

Introduction

Brain abscess in adults arises mostly due to hematogenous spread of bacteria from foci of lung abscesses, bronchiectases or empyema. Other common etiologic factors include contiguous spread from purulent sinusitis and middle ear infection as well as infections following penetrating cranial trauma or neurosurgical procedure [5]. Pulmonary arteriovenous malformation (PAVM) can also lead to the formation of a cerebral abscess [3,7]. Despite the fact that cases of PAVM occur very seldom (more than 500 cases reported in the literature), PAVMs are common in patients suffering from hereditary hemorrhagic telangiectasia (HHT, Rendu-Osler-Weber disease) [3,11]. Typically PAVMs tend to remain asymptomatic and neurological complications, the most serious of which is a brain abscess, can be the first clinical manifestation of PAVM.

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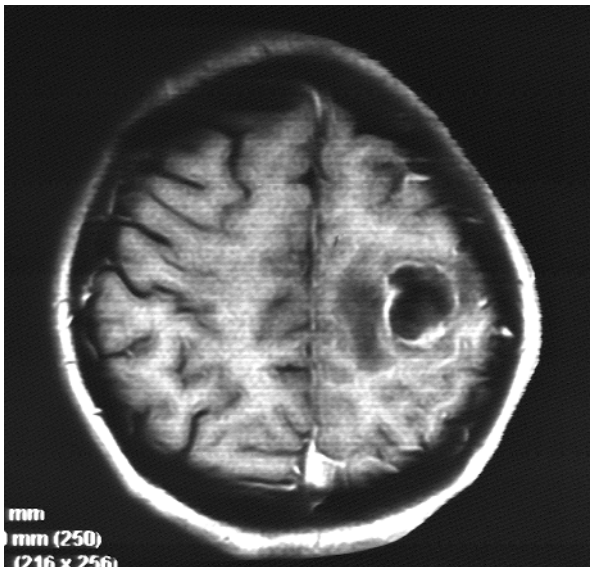

Fig. 1. MRI scan showing a cystic tumor in the left parietal lobe with irregular ring enhancement. The tumor is surrounded by brain edema

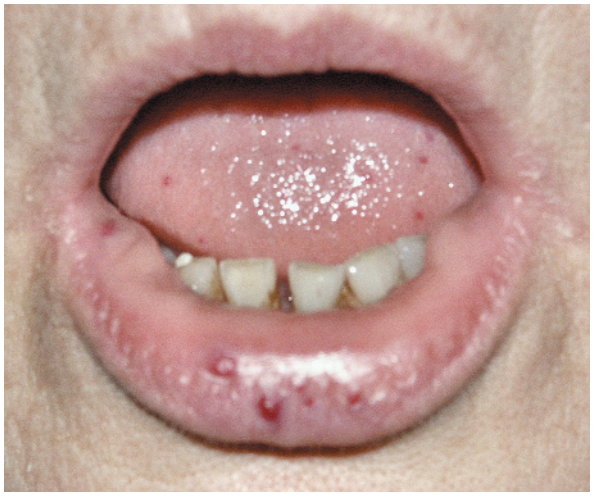

Fig. 2. Photograph presenting multiple small raised purple telangiectases on the lips and the tongue

Case presentation

A 53-year-old woman was admitted on April 02 2004 to the Department of Neurology of the District Hospital in Jelenia Gora because of acute onset of headache, nausea and vomiting, accompanied by increasing right hemiparesis, motor aphasia and gait disturbances. Computed tomography (CT) of the head and later magnetic resonance investigation (MRI) revealed a left parietal cystic tumor, surrounded by cerebral edema, showing ring enhancement following contrast administration (Fig. 1) It looked more than an abscess but a neoplastic tumor could not be excluded at that time. The patient was qualified for operative treatment and transferred to the Department of Neurosurgery Wroclaw Medical University on April 09 2004.

Examination. On admission the woman was lethargic and disoriented. She presented moderate right hemiparesis, more prominent in the upper limb, right-side facial weakness and motor aphasia. Babinski's sign was present on the right. The patient was afebrile and without evidence of dyspnea, cyanosis, tachypnea or digital clubbing in the hands and feet. Yet physical examination revealed signs of systemic anemia as well as multiple small raised cutaneous and mucosal telangiectases, present on the fingertips, lips and on the oral cavity – and nasal mucosa (Fig. 2). Past medical history indicated that the patient had been treated for 15 years because of recurrent epistaxis and iron deficiency anemia.

Laboratory evaluation was notable for a white blood cell count of 20.710/µl with normal differential counts, hemoglobin 7.9 g/dl and erythrocyte sedimentation rate 30 mm/hr. Arterial blood gas analysis was normal (PaO_2 83.6 mmHg, $PaCO_2$ 36.1 mmHg, HCO_3 25.8 mmol/l, Sat O_2 97%, pH 7.46).

Basing on the history of recurrent epistaxis as well as the presence of cutaneous and mucosal telangiectases, we set the initial diagnosis of hereditary hemorrhagic telangiectasia. In light of this finding, it was more likely that the parietal tumor was an abscess.

Operation. The tumor mass was exposed following a left parietal craniotomy. It was situated in the superficial layers of the parietal lobe, occupying a 2.5 cm^2-area of the precentral gyrus. The tumor was encapsulated, well demarcated and had the appearance of an abscess. The abscess was drained with a brain needle and 10 ml of pus was aspirated.

Postoperative course. The patient was treated for 6 weeks with intravenous administration of ceftriaxone and amicacin. Culture of the pus obtained intraoperatively was negative. The patient's neurological condition improved with the management. Symptoms of elevated intracranial pressure, aphasia

and gait disturbances disappeared. The right hemiparesis markedly decreased. Only a discrete weakness of the elbow and wrist flexors was observed. Brain CT obtained 5 weeks following the operation showed only residual areas of brain edema around the abscess cavity, without any mass effect.

Taking into consideration our initial diagnosis of HHT, it was now easier for us to determine the origin of the brain abscess. The contrast-media-enhanced chest CT study revealed 6 arteriovenous fistulae, localized in the lower lobe of the right lung as well as gave evidence of advanced pulmonary embolism (Fig. 3, Fig. 4). Since most of the PAVMs had a feeding artery exceeding 3 mm in diameter, the patient was qualified for pulmonary angiography and subsequent embolotherapy.

CT scans of the abdomen did not reveal any vascular malformations in the liver. Gastroscopy was also negative. An analysis of the preoperative MRI scans excluded the existence of cerebral arteriovenous malformations.

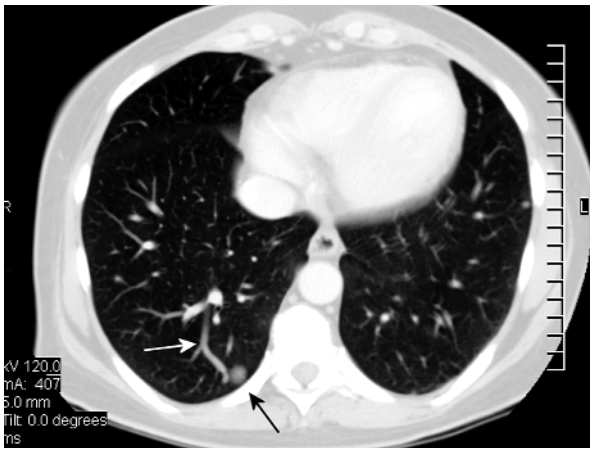

Fig. 3. Contrast-media-enhanced chest CT scan. Axial plain reveals one of the six arteriovenous fistulae, present paravertebrally in the lower lobe of the right lung (black arrow). The white arrow shows the segmental artery feeding the fistula

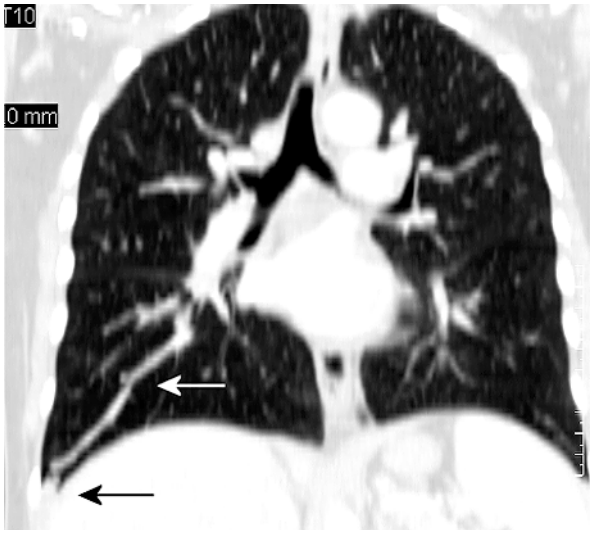

Fig. 4. Frontal reconstruction of the chest CT. An arteriovenous fistula is shown just above the diaphragm (black arrow). The fistula is fed by a segmental artery exceeding 3 mm in diameter (white arrow)

Discussion

The cerebral abscess we diagnosed and treated, developed in a patient suffering from hereditary hemorrhagic telangiectasia (HHT, Rendu-Osler-Weber disease). It is a rare autosomal dominant disease, characterized by episodes of spontaneous and recurrent epistaxis, cutaneous and mucosal telangiectases and visceral arteriovenous malformations. The visceral vascular anomalies include pulmonary, hepatic and cerebral arteriovenous fistulae and telangiectases in the esophageal, gastric and intestinal mucosa. Recently, in order to make the diagnosis of HHT easier, Shovlin et al. proposed the following diagnostic criteria: patient's familiar history, epistaxis, telangiectases and visceral arteriovenous malformations [10]. The diagnosis of HHT is certain if 3 out of 4 criteria are satisfied and suspected if 2 out of 4 criteria are satisfied. Our patient meets the criteria of HHT because of history of recurrent epistaxis as well as the presence of cutaneous and mucosal telangiectases and multiple PAVMs in the right lung. Clinical observations show that 15% of the patients with HHT have PAVM [6,11]. PAVM is characterized by a pathological connection between an afferent artery and one or more efferent veins without an interposed capillary bed [9]. PAVMs remain for many years unrecognized, as shown in this case. According to Press and Ramsey, signs of hypoxemia like dyspnea, cyanosis, polycythemia and clubbing of the fingers and toes are observed only in one-third of the cases of PAVM [8]. The asymptomatic character of PAVMs does not mean that they are inactive. In fact, right-to-left shunts in the lung circulation, formed by PAVMs, lead to chronic

hypoxemia and secondary polycythemia [1]. With time thromboemboli arise within the pulmonary fistula. In addition, right-to-left extracardiac shunt stimulates the proliferation of anaerobic bacteria. Thromboemboli and septic microemboli, arising in the pulmonary circulation, evoke cerebrovascular disorders. Adams et al. reported that transient ischemic attacks and lacunar brain strokes are usually the first clinical manifestation of pulmonary embolism resulting from PAVM [1]. Yet a brain abscess is the most serious neurological complication of PAVM and occurs in 5% to 10% of patients with PAVM [6]. It is believed that right to left shunting in the lungs enables bacteria to avoid the filtering effect of pulmonary capillaries. In consequence, the brain becomes the first and the most frequently affected target for bacterial emboli [1,9]. Our observation is consistent with the experience of other neurosurgeons that a brain abscess may be the first clinical manifestation of an asymptomatic PAVM [6,7]. Besides, a PAVM should be considered in each case of a brain abscess of unknown etiology.

Several characteristics of cerebral abscesses may indicate their attribution to PAVM. Firstly, they develop often during the third to fifth decade of life, when PAVMs increase in size and number [1]. Typically these abscesses are multiple and recurrent [9]. Distinct from other hematogenous brain abscesses, PAVM-evoked brain abscesses affect the superficial layers of the cerebral lobes (mostly the parietal lobe), where microinfarction and septic embolization often occur [3,9]. Furthermore, a brain abscess in a patient with HHT increases the probability of PAVM to 90% [6,7]. As illustrated by this case, most of the mentioned features of a cerebral abscess associated with PAVM could be observed.

Treatment of a brain abscess in patients with PAVM requires firstly aspiration or excision of the abscess and then embolization of the PAVM. We claim that chest-CT should be the screening investigation for PAVM. Each PAVM with feeding artery exceeding 3mm is currently recommended for embolotherapy [2,12]. Pulmonary angiography should precede the embolization [6].

Patients suffering from HHT belong to the group of high risk of development of PAVM, which if not treated on time may cause a brain abscess. Patients with diagnosed HHT as well as their family members, suspected to have HHT, should undergo a complete diagnostic study, aimed at elucidation of visceral vascular malformations.

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