Large Pulmonary Arteriovenous Malformation with Hyperammonemia

A 45-year-old female presented with generalized fatigue, unaccompanied by other symptoms. Investigation revealed severe anemia due to gastric bleeding, and hereditary hemorrhagic telangiectasia accompanied by a large pulmonary arteriovenous malformation (PAVM). Additionally, the presence of hepatic arteriovenous and portovenous shunts indicated hepatic involvement. In addition to hypoxemia due to right-to-left shunting in PAVM, hyperammonemia with normal hepatic function was detected. The large PAVM was successfully managed with surgical resection. Hyperammonemia, persisting despite the hemostasis of gastric bleeding, improved postoperatively in the absence of treatment directed at hepatic involvement. We believe that resection of large PAVM contributed to the improvement of hyperammonemia. (Jpn J Thorac Cardiovasc Surg 2004; 52: 484–487)

Key words: hereditary hemorrhagic telangiectasia, pulmonary arteriovenous malformations, hyperammonemia

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HEREDITARY HEMORRHAGIC TELANGIECTASIA (HHT) is an autosomal dominant disease characterized by abnormal vascular structures affecting many organs, including the skin, lungs, gastrointestinal tract, brain and liver. Pulmonary arteriovenous malformations (PAVM) are rare, mostly congenital, abnormalities often associated with HHT. They cause right-to-left shunts leading to hypoxemia, which can result in more serious complications such as hemoptysis, hemothorax and systemic embolism. Therefore, even if patients are asymptomatic, appropriate treatment is strongly recommended. Hepatic vascular anomalies are also found in some patients affected by HHT and can cause complications. We herein report a case of HHT associated with large PAVM, hepatic involvement, and hyperammonemia, treated by surgical resection.

Case

A 45-year-old female presented with generalized fatigue in August 2002. No medical history of note was obtained, with epistaxis in particular being denied. On examination, a systolic murmur was audible over the left anterior chest. Investigations revealed severe anemia (Hb 2.8 mg/dl, microcytic); however, no abnormalities in urinalysis or blood chemistry were noted. Gastrointestinal fiberscopy (GIF) demonstrated bleeding from a gastric ulcer in addition to gastric telangiectasia. Chest radiography showed a sharply defined multinodular lesion, 69x23 mm in diameter, in the left lower lung field (Fig. 1).

Blood transfusion was required to correct the anemia and gastric bleeding was treated medically using a proton pump inhibitor. After recovery from generalized fatigue and anemia, hypoxemia was revealed (pH 7.495, PaO2 62.1 mmHg, and PaCO2 22.3 mmHg on room air), although the patient did not describe symptoms of dyspnea or cyanosis.

Contrast-enhanced computed tomography (CT) demonstrated the pulmonary lesion to be a vascular abnormality. Pulmonary angiography displayed a large PAVM, 40x57 mm in diameter, fed by the left pulmonary arteries A4 and A5 (Fig. 1). The diameters of the feeding arteries were measured at 13 mm and 10
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A: Chest radiograph revealing a round mass in the left lung.
B: Pulmonary angiogram showing a big pulmonary arteriovenous malformation fed by 2 large pulmonary arteries.

Cerebral magnetic resonance imaging (MRI) revealed multiple small lacunar infarctions despite no episodes of neurologic complications. Abdominal CT demonstrated hepatic artery dilatation and disseminated peripheral telangiectasia in the liver (Fig. 2). Hepatic complications, such as arteriovenous, arteriportal or portovenous shunting, were therefore suspected. Moreover, an elevated level of ammonia (115 μmol/l) was detected despite hemostasis of gastric bleeding.

GIF revealed some remaining gastric telangiectasia. A clinical diagnosis of HHT was made while no positive family history nor genetic consultations were obtained.

For the purposes of preventing complications of HHT and improving gas exchange, lingular segmentectomy was performed via left thoracotomy in order to resect the PAVM. Postoperatively, the bruit over the left anterior chest wall resolved and PaO₂ improved (pH 7.474, PaO₂ 87.4 mmHg, PaCO₂ 30.7 mmHg on room air). Furthermore, although no treatment targeted at the hepatic involvement was administered, the serum level of ammonia fell to 51 μmol/l. Postoperative pulmonary angiography detected no residual PAVM and the patient remains in good health at the present time, with no symptoms related to HHT in evidence.

Discussion

Patients with HHT have a high incidence of telangiectasia in multiple organs, with the earliest and most common symptom being epistaxis, caused by bleeding from mucosal telangiectasia. The incidence of bleeding from telangiectasia in the gastrointestinal tract in patients with HHT is 20%, and severe bleeding may lead to