Red Papules on the Tongue of a Patient With Hemiparesis

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A 39-year-old woman had an acute onset of headache, increasing right-side hemiparesis, and numbness with fever. Computed tomography (CT) of the brain showed a ring-enhanced lesion with surrounding edema on the left frontoparietal lobe. Multiple red papules on the tongue (Figure 1), trunk, and limbs were noted during the physical examination. The patient stated she had not noticed the red papules before. There was no suggestion of otitis media or paranasitis and no history of craniotomy, open head injury, or cyanotic heart disease. The patient did relate having many prior episodes of epistaxis since childhood. Blood cultures were obtained when the patient was first seen, and the patient received ampicillin/sulbactam empirically.

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WHAT WOULD YOU DO NEXT?

A. Consult a neurosurgeon to evaluate the brain CT finding; obtain a CT scan of the lung and abdomen and offer genetic counseling for the patient and her family.

B. Consult a neurosurgeon to evaluate the brain CT finding; obtain a dermatology consultation and refer the patient to interventional radiology.

C. Continue intravenous antibiotics only.

D. Obtain a biopsy of the tongue lesions.
Diagnosis
Rendu-Osler-Weber syndrome (hereditary hemorrhagic telangiectasia [HHT]) presenting with a brain abscess.

What To Do Next
A. Consult a neurosurgeon to evaluate the brain CT finding; obtain a CT scan of the lung and abdomen and offer genetic counseling for the patient and her family.

Red telangiectasias on the tongue and a history of recurrent epistaxis suggest HHT. Brain abscesses can occur in HHT because of arteriovenous shunting. A search should thus be made for arteriovenous malformations of the lung and liver by obtaining CT scans of the chest and abdomen. Queries should be made regarding a family history of bleeding or lesions characteristic of HHT.

Discussion
Rendu-Osler-Weber syndrome (HHT) is an autosomal dominant vascular dysplasia affecting multiple systems.1,2 The syndrome is characterized by epistaxis, cutaneous telangiectasia, and visceral arteriovenous malformations. Most of the cutaneous telangiectasia develops by age 40 years, usually before the appearance of pulmonary arteriovenous malformation-associated complications, such as brain abscess.3 Pulmonary arteriovenous malformation occurs in 5% to 30% of patients with HHT.4,14 The greatest morbidity is development of pulmonary arteriovenous malformations and neurologic complications.1,5 Emboli may cause transient ischemic attacks, infarcts, and seizures, whereas septic emboli cause brain abscesses.1,6,15 Brain abscesses in patients with HHT have concomitant pulmonary arteriovenous malformations in 90% of cases.16 Brain abscess attributable to vascular shunting of pulmonary arteriovenous malformations can often be the first manifestation, because 75% of pulmonary arteriovenous malformations in patients with HHT are asymptomatic.6

The differential diagnosis of cerebral ring-enhancing lesions includes abscess, subacute infarction or hemorrhage, demyelinating diseases, and neoplasms. Pyogenic abscesses remain the most likely diagnoses, particularly in this febrile patient. Physicians evaluating patients with brain abscess should look for the primary source of infection, from direct extension of a contiguous suppurative focus, from hematogenous dissemination of emboli, or from direct inoculation associated with trauma or neurosurgical procedures. If none of these are present, right-to-left shunts from congenital heart diseases or arteriovenous malformations should be considered. Up to 5% of patients with pulmonary arteriovenous malformations develop brain abscesses.7 About 60% to 90% of patients with such malformations have HHT.6 Red tongue nodules occur in Kawasaki disease (strawberry tongue), blue rubber bleb nevus syndrome,8 scarlet fever, lichen planus, syphilis (tongue sore), and herpetiform glossitis. However, having these lesions along with brain abscess and a history of recurrent epistaxis suggests HHT. A diagnosis of HHT is considered definite if 3 or more of the following “Curaçao” criteria are met: (1) epistaxis; (2) cutaneous and mucosal telangiectasias; (3) visceral arteriovenous malformation; and (4) a family history in which a first-degree relative meets these criteria.8 Obtaining a tongue biopsy would have shown telangiectasia and would help establish a diagnosis of HHT based on Curaçao criteria. However, finding a multiplicity of arteriovenous malformations on CT can also be diagnostic for HHT without requiring an invasive procedure such as a biopsy.

Treatment for HHT varies by the involved site and how the disease presents. Cautery, sclerotherapy, embolization, endoscopic laser therapy, and surgical resection have been used.3 Bevacizumab, a vascular endothelial growth factor inhibitor, has shown promise in reducing shunting and its complications in HHT.9,10 Major fatal complications of HHT include uncontrolled epistaxis, gastrointestinal hemorrhage, stroke, deep venous thromboses, severe hepatic shunting causing heart failure, and severe pulmonary shunting with hypertension.9

Patient Outcome
The brain CT showed an abscess, which was drained. Cultures grew Actinomyces meyeri. Chest CT and angiography demonstrated multiple arteriovenous fistulae located predominantly in the bilateral lower lung lobes. Intrahepatic vascular malformation was seen on abdominal CT. The patient’s neurologic symptoms improved following 4 weeks of ampicillin/sulbactam, penicillin G, and gentamicin. Further exploration of the patient’s family history revealed that 9 family members in 3 generations had recurrent epistaxis, pulmonary arteriovenous malformation, or both, possibly caused by HHT.

REFERENCES