Clinical Reasoning: A rare cause of subarachnoid hemorrhage

A 48-year-old woman presented with severe headache radiating to her neck and chest, followed by a brief period of loss of consciousness in the emergency department. After she regained consciousness, the patient described a 2-year history of right-sided pulsatile tinnitus and hearing loss. She also had a history of poorly controlled hypertension and of noncompliance with her medication. On examination her blood pressure was 239/90 mm Hg. Results of a neurologic examination were unremarkable. Brain CT showed a subarachnoid hemorrhage (figure 1, A and B).

Questions for consideration:
1. What are the possible etiologies of her subarachnoid hemorrhage?
2. What additional diagnostic testing would you consider at this point?

Figure 1 Initial head CT scan revealing subarachnoid hemorrhage

(A) Noncontrast brain CT scan demonstrates a posterior fossa mass and subarachnoid hemorrhage. (B) Intraventricular hemorrhage is also demonstrated.

SECTION 1

A 48-year-old woman presented with severe headache radiating to her neck and chest, followed by a brief period of loss of consciousness in the emergency department. After she regained consciousness, the patient described a 2-year history of right-sided pulsatile tinnitus and hearing loss. She also had a history of poorly controlled hypertension and of noncompliance with her medication. On examination her blood pressure was 239/90 mm Hg. Results of a neurologic examination were unremarkable. Brain CT showed a subarachnoid hemorrhage (figure 1, A and B).

Questions for consideration:
1. What are the possible etiologies of her subarachnoid hemorrhage?
2. What additional diagnostic testing would you consider at this point?
CT angiography of the head and neck did not reveal any intracranial aneurysm or vascular malformation (figure 2, A and B). However, a large mass was present in the right jugular foramen with erosion of temporal bone and encroachment of the right internal acoustic canal. MRI confirmed a hypervascular mass that contained numerous flow voids. The mass was located lateral to the medulla and extended superiorly into the cerebellopontine angle cistern. It extended inferiorly to the superior aspect of the right parapharyngeal space and displaced the right internal carotid artery (figure 2C).

Catheter-based cerebral angiography demonstrated an intense hypervascular tumor blush extending from the superior aspect of the right carotid sheath into the skull base (figure 2D). The major feeding arteries were from branches of the right internal maxillary artery, ascending pharyngeal artery, and posterior auricular artery. Ear, nose, and throat evaluation revealed a reddish mass in the middle ear visible through the tympanic membrane and right vocal cord paresis with minimal abduction. The patient denied any hoarseness, dysphagia, dysarthria, or focal neurologic deficits.

Questions for consideration:
1. What is your differential diagnosis at this point?
2. How would you manage this patient?

(A) CT angiogram showing an enhancing jugular foramen mass extending into the posterior fossa. (B) Enhancing tumor mass at the skull base. (C) MRI T2-weighted image showed salt-and-pepper appearance. (D) Selective right ascending pharyngeal angiogram lateral view shows a hypervascularized tumor blush, fed by 2 major branches from the enlarged right ascending pharyngeal artery.
A 24-hour urine sample demonstrated elevated meta-nephrine (1,519 μg/24 h; reference, 95–475 μg/24 h) and normetanephrine (567 μg/24 h; reference, 52–310 μg/24 h) levels but normal urinary vanillylmandelic acid. There was no adrenal mass on abdominal MRI. The patient’s hypertension was treated with phenoxybenzamine, nicardipine, metoprolol, and hydralazine.

After preoperative embolization of the right external carotid artery feeders, the patient underwent tumor removal by a transpetrosal/infratemporal fossa approach. A large red mass was seen within the middle ear space with substantial extension through the skull base and intracranially. The tumor extended through the jugular foramen, and there was considerable erosion of the petrous apex of the temporal bone. The intracranial tumor abutted the lateral portion of the lower pons and medulla. There were several large arterial feeders from the anterior inferior cerebellar artery. The internal jugular vein was massively distended with tumor extending several centimeters inferiorly inside the lumen of the vein. This portion of the tumor was removed en bloc and measured 8.0 cm by 1.7 cm by 0.8 cm. It was a sausage-shaped hemorrhagic tumor with gelatinous appearance, which on cut surface examination had a homogeneous tan pinkish appearance (figure 3, A and B). The remainder of the tumor was removed in a piecemeal fashion. Access to the tumor required rerouting of the facial nerve. The tumor displaced the nearby glossopharyngeal, vagus, spinal accessory, and hypoglossal nerves.

Microscopically, the tumor was very vascular (figure 3, C and D). The vessels within the tumor separated the cells into irregular groups causing a zellballen pattern (figure 3, C and D). The vessels were distinctly stained by CD34 (figure 3D). The chief cells were positive by chromogranin (figure 3E) and synaptophysin (not shown). The sustentacular cells were positive by S100 (figure 3F). The anatomic diagnosis was jugulotympanic paraganglioma. Immediately after surgery, her tinnitus resolved. The patient developed new facial weakness from rerouting of the facial nerve, which continues to improve. Her hypertension has also improved but has not completely resolved.

**DISCUSSION** A paraganglioma is a very unusual cause of subarachnoid hemorrhage. To our knowledge, this is the second case reported in the literature. Paragangliomas, also known as chemodectomas or glomus tumors, are rare tumors derived from the extra-adrenal paraganglionic tissue that is thought to originate from the neural crest. They are biologically similar to pheochromocytomas. It has been estimated that paragangliomas comprise 1 in 30,000 of all head and neck tumors. Paragangliomas of the head and neck are seen in 4 primary locations: the jugular bulb, the middle ear cavity, the vagus nerve, and the carotid body. The most common paraganglioma of the head and neck is the carotid body tumor, followed by jugulotympanic paragangliomas (glomus tympanicum and glomus jugulare) and then by vagal paragangliomas (glomus vagale). Glomus tympanicum is the most common neoplasm of the middle ear and arises from the paraganglionic tissue in the cochlear promontory. Glomus jugulare tumors arise from paraganglionic tissue along the jugular bulb adventitia. When their origin to the promontory cannot be ascertained, they are called jugulotympanic paragangliomas.

Histologically, paraganglia contain 2 cell types: chief cells and supporting sustentacular cells. These
Radiotherapy or stereotactic radiosurgery can be used as an adjunct to surgery and as a primary treatment for healthy patients with functional cranial nerve deficits. At an earlier stage at diagnosis, and difficult anatomic location.

Lower cranial neuropathies (glossopharyngeal, vagus, accessory nerves) are related to the growth of this very vascular tumor within the middle ear space. At later stages, which are related to the growth of this very vascular tumor, lower cranial neuropathies (glossopharyngeal, vagus, accessory nerves) are also common. Clinically significant hormone secretion is present in only 2% of patients. Symptoms suggestive of hormonally active tumors (e.g., labile hypertension, facial flushing, and palpitations) mandate a thorough evaluation to rule out pheochromocytoma.

Asymptomatic, is characteristic of these tumors. Neuroimaging is important for evaluating tumor vascularity and extent. On T1-weighted images pheochromocytomas appear hypointense with a speckled appearance. On gadolinium-enhanced T1 images, early and pronounced enhancement is seen, witnessing the hypercellular nature of these lesions and their frequent association with hypertension. The most common diagnosis suggested was a pheochromocytoma, followed by a paraganglioma.

CT imaging is excellent for demonstrating characteristic bone destruction and hyperdense calcifications, and it is superior for evaluating tumor vascularity and extent. MRI is the diagnostic modality in patients who are poor surgical candidates or those with bilateral disease.3

Paragangliomas are considered benign but can cause extensive destruction by their unrestrained growth. Patients with glomus jugulare often present with similar clinical features such as pulsatile tinnitus and glomus jugulare often present with similar clinical features.
50% of respondents. Additional considerations, in descending order of the number of times that they were cited, included arteriovenous malformation, cerebellopontine angle tumor, carotid dissection, congenital malformation, and reversible cerebral vasconstriction syndrome.

The most frequently recommended diagnostic test was angiography (54% of respondents). The other recommendations in descending order of the number of times that they were cited included MRI with contrast, 24-hour urine collection for catecholamine metabolites, otoscopy, and CT of the chest, abdomen, and pelvis.

This is the third Mystery Case published in the Resident & Fellow section of Neurology®. This case demonstrates the importance of multimodality imaging and attention to systemic symptoms, which may aid the practitioner in making the correct diagnosis.

Keith R. Ridel, MD
Clinical Reasoning: A rare cause of subarachnoid hemorrhage
A. Emami, K. Panichpisal, E. Benardete, et al.
Neurology 2011;76:e43-e47
DOI 10.1212/WNL.0b013e3182104330

This information is current as of March 14, 2011