Carotid body tumor as a potential cause of paroxysmal hemicrania

Caywin Zhuang, BA,[a] *and* Brian Beeman, MD,[b] *Champaign and Urbana, Ill*

ABSTRACT

A 70-year-old woman presented to our emergency department with a severe left-sided headache. The headache's location and accompanying symptoms are consistent with paroxysmal hemicrania. On workup, a cervical computed tomography scan incidentally revealed a left carotid body tumor, and the patient was referred to vascular surgery. After the resection of the tumor, the patient recovered well in the following months. Furthermore, she no longer experienced any additional headache episodes, which were likely caused by the carotid body tumor. (J Vasc Surg Cases and Innovative Techniques 2020;6:495-7.)

Keywords: CBT; Paraganglioma; PH; Headache; Vascular surgery

Paragangliomas are rare neuroendocrine tumors derived from the embryonic neural crest and have the ability to secrete catecholamines. They arise most commonly from the carotid body, which are then referred to as carotid body tumors (CBT). CBTs are usually asymptomatic and discovered as a painless mass of the neck at the level of the carotid bifurcation. Larger CBTs can sometimes be associated with symptoms of a space-occupying lesion in this location. Rare functional CBTs may cause sympathetic hypersensitivity, presenting as palpitations, hypertension, tachycardia, and headache from the release of catecholamines. However, the clinical presentation of secondary paroxysmal hemicrania (PH), which resolves after surgical removal of a nonfunctional CBT, constitutes a unique case. Consent for publication has been obtained from the patient.

CASE REPORT

A 70-year-old woman presented to the Carle Emergency Department with a 2-week history of intractable left-sided headache. She described her headache as a throbbing, sharp pain that begins above her left eye and radiates to the left parietal area. She came to the hospital owing to the headache's worsening severity refractory to acetaminophen. Associated symptoms included photophobia, lacrimation, positional dizziness, and occasional ear pain. She was a former smoker with a history of chronic obstructive pulmonary disease, hypertension, myocardial infarction, left bundle branch block, and implantable cardioverter-defibrillator in place. She did not have a history of headaches. On physical exam, some fullness was noted in the left neck, with no carotid bruits bilaterally. A computed tomography (CT) angiogram with contrast revealed an avidly enhancing mass within the left carotid sheath measuring 10 mm × 15 mm × 14 mm (Fig 1). The mass splayed the left proximal internal and external carotid arteries. Serum and urine metanephrine results were negative. These findings were consistent with a nonfunctional CBT, preoperatively classified as Shamblin type I. Neurology ruled out intracranial causes, and the patient was started on indomethacin.

Given a potential link to the patient's headache, vascular surgery was consulted and patient underwent surgical resection of the tumor. A small CBT was dissected free from the bifurcation of the internal and external carotid arteries (Fig 2). After the surgery, the patient showed a transient nerve palsy marked by numbness and drooping in the left lip, which had resolved by her 2-month follow-up. In addition, she stated that her headache, photophobia, and other associated symptoms have been completely resolved, with no relapse or need for medications in the 7 months after discharge.

A pathology study confirmed the benign nature of her carotid body paraganglioma (Fig 3).

DISCUSSION

Paragangliomas develop from the parasympathetic system in the head and neck, with 60% of these tumors occurring in the carotid body.[1] Paragangliomas may be hereditary or part of genetic syndromes such as Von Hippel-Lindau syndrome, neurofibromatosis type 1, and multiple endocrine neoplasia type 2.[2] Hereditary cases are linked to alterations to the succinate dehydrogenase family of genes.[1] However, even with modern genetic testing, most paragangliomas appear to be sporadic.[3]

Sporadic CBTs are more prevalent among women.[4] They are also associated with chronic hypoxia as a risk factor, frequently found in patients living at high altitudes.[5] Although the causative role of chronic hypoxia remains speculative, the carotid body is known for its adaptive function to fluctuating oxygen, carbon

From the Carle Illinois College of Medicine, Champaign[a]; and the Carle Heart and Vascular Institute, Urbana.[b]

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Correspondence: Caywin Zhuang, BA, Carle Illinois College of Medicine, 807 S Wright St, Champaign, IL 61820 (e-mail: cqz2@illinois.edu).

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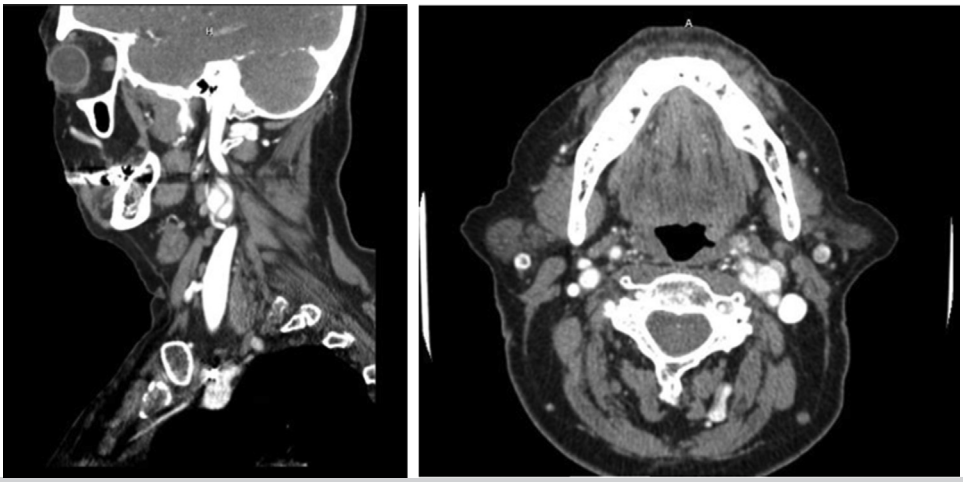

Fig 1. Sagittal (*left*) and axial (*right*) computed tomography (CT) angiography of the neck with contrast showing intense enhancement of the carotid body tumor (CBT).

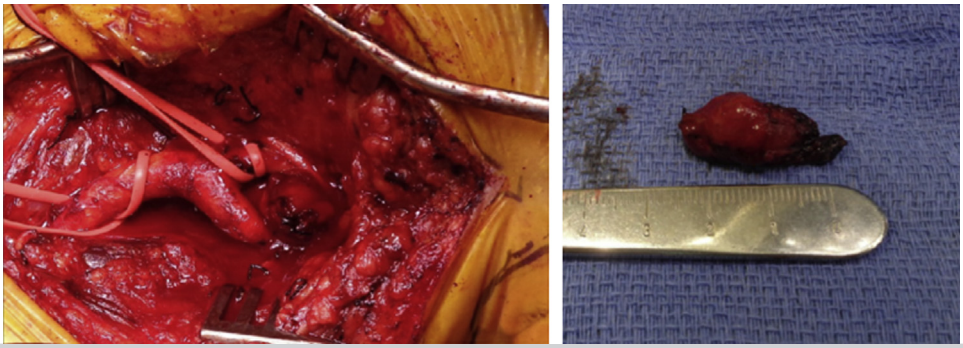

Fig 2. Intraoperative pictures before and after resecting the carotid body tumor (CBT), measuring 23 mm in length.

dioxide, and proton concentration. Therefore, chronic hypoxic stimulation may cause carotid body hypertrophy, ultimately leading to pathologic hyperplasia and/or neoplastic development.[6] Our patient has a history of chronic obstructive pulmonary disease and no known familial syndromes. Her hypoxic respiratory condition has likely contributed to the development of a sporadic CBT. Even in seemingly sporadic cases such as this one, some studies have also recommended succinate dehydrogenase mutation screening for patient and family for early detection of synchronous lesions.[7-9]

The majority of CBTs are asymptomatic and initially noticed as neck swelling during a physical examination or, more commonly, as incidental findings on imaging studies.[9-11] Very large tumors may cause compression or local invasion, leading to nonspecific symptoms such as localized tenderness, fullness, numbness, dysphagia, hoarseness, chronic cough, and tinnitus. In rare cases of functional tumors, neuroendocrine activity may lead to complaints of headaches, dizziness, palpitations, tachycardia, arrhythmias, flushing, diaphoresis, and photophobia. Some cases have reported hypertension that resolves after tumor resection, which may be caused by tumor-related catecholamine release.[12]

Although this patient's symptoms of headache and hypertension may suggest a functional CBT, her serum catecholamine levels were within normal range. Rather than sympathetic hyperactivity, her chief complaint of headache is more consistent with a diagnosis of secondary PH. PH can present similarly as cluster headache, but it differs in a predominance in females[13] and responsiveness to indomethacin.[14] This latter differentiation can be clinically useful for treatment selection if her headache had persisted after tumor resection. The headache of PH is strictly unilateral. The pain is most often in the ophthalmic trigeminal distribution, but it can be extratrigeminal. The maximum pain is usually centered on the orbital, retroorbital, temporal, and frontal regions.[15] In addition, photophobia and nausea may accompany some attacks,[13] as seen in the present case. PH may also be secondary to other intracranial and vascular pathologies, which were less likely given her unremarkable head CT scan.

The pathophysiology of PH is largely unknown, but the present case helps to establish CBT as a potential trigger.

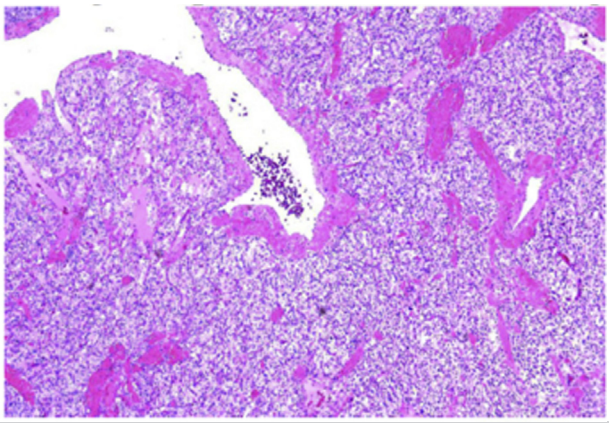

Fig 3. Histologic analysis shows nests of tumor cells showing moderate to abundant amphophilic cytoplasm separated by sustentacular cells. No malignancy is seen.

Although it is difficult to prove absolute causation, we report at least a temporal connection between the tumor removal and headache relief in this patient. Her presentation is consistent with the hypothesis of a cervicogenic origin of PH, based on studies showing a clear temporal association of symptoms with neck movement.[16] The onset of her headache indicates a mechanical precipitation mechanism located somewhere in the neck.

Surgical resection is the treatment of choice for CBT.[9-11] It can be guided by a classification system developed by Shamblin et al[17] to better predict surgical morbidity. It divides the tumors into three groups based on tumor extent and neurovascular involvement. We did not preoperatively embolize this small, Shamblin type I CBT. It was not adherent to the adventitia and did not surround the carotid vessels; thus, the risk of inadvertent cerebral embolization and stroke was deemed too high. Recent study has also demonstrated tumor distance to base of skull and tumor volume as quantifiable predictors of bleeding and cranial nerve injury risk.[18] Her tumor volume was 1.1 cm^3 based on dimensions measured by CT angiography. Located 4.3 cm distal to the skull base, it was readily accessible by standard neck incision. These measures predicted low operative risks, consistent with the surgical outcome.

Modern imaging allows staging of CBTs according to the Shamblin classification and can assist in preoperative planning. Axial imaging, either CT angiography or magnetic resonance imaging is the preferred modality because it best defines the relationship of the tumor with the artery bifurcation and the likely location of the cranial nerves.[19] Only CT imaging was performed in this patient because her pacemaker precluded her from magnetic resonance imaging. However, incidental findings of the classic radiographic features of a CBT, which has likely triggered her headache, demonstrates the diagnostic value of this workup.

To conclude, CBT is a rare lesion that can be difficult to diagnose, especially with atypical presentations like PH shown in the current clinical case. Imaging studies are useful if CBTs are suspected in the differentials of headaches.

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