Diagnosis and Management of Cervical Sympathetic Chain Paraganglioma

Rahul Seth MD1, Karah M. Lanier MD2, Benjamin G. Wood MD3, Aaron P. Hoschar MD3, Joseph Scharpf MD1

1Head and Neck Institute, 2Radiological Institute, 3Pathology and Laboratory Medicine Institute

Cleveland Clinic, Cleveland, Ohio

ABSTRACT

OBJECTIVE: At the conclusion of this presentation, the participant should be able to identify that cervical sympathetic chain paragangliomas are a unique entity with distinct clinical presentation, imaging characteristics, and management.

INTRODUCTION: Paragangliomas are rare, slow-growing, neuroendocrine tumors that develop at autonomic ganglia. In the head and neck region, they most commonly occur at the carotid body, jugular bulb, and vagus nerve. Paragangliomas arising from the cervical sympathetic chain are exceedingly rare, with only nine cases reported in the literature.

STUDY DESIGN: A retrospective case study of two patients and review of the literature.

METHODS: We describe the presentation, imaging, and management of cervical sympathetic chain paragangliomas in two patients. One patient presented with ipsilateral Horner’s syndrome, and both had posterior pharyngeal submucosal masses. MRI characteristics were consistent with paragangliomas with retropharyngeal space and anterolateral displacement of both the carotid and jugular vessels.

RESULTS: Intraoperatively, both tumors arose from the cervical sympathetic chain and were hypervascular. Histopathological analysis confirmed diagnosis of paraganglioma.

CONCLUSIONS: Paragangliomas arising from the cervical sympathetic chain are exceptionally rare but must remain in the differential diagnosis of paragangliomas masses. They may present with ipsilateral Horner’s syndrome and posterior pharyngeal fullness. Imaging characteristics are typical for paragangliomas with anterolateral displacement of both the carotid and jugular vessels.

CASE 1

A 33 year old woman presented with complaint of new onset hyponasal voice, nocturnal snoring, and posterior oropharyngeal fullness causing mild dysphagia. She denied dyspnea, palpitations, hypertension, tachycardia, weight loss, and night sweats. She had no significant medical history, and no family history of paraganglioma.

Physical examination revealed a right posterior oropharyngeal submucosal mass that extended to the left of the midline. Flexible nasopharyngoscopy revealed the submucosal mass to extend superiorly to the right nasopharynx along the lateral pharyngeal wall. There was no palpable neck mass or fullness. She had no anisocoria or ptosis.

Contrast CT showed intensely enhancing 3.9 x 5.9 cm mass in right parapharyngeal space extending from the medial carotid sheath to the oropharyngeal wall. MRI with contrast revealed similar findings with flow voids on T2 imaging (Figure 1). Octreotide scan was positive for this lesion and indicated that it was resectable.

Post-operatively she developed an expected Horner’s syndrome and first bite syndrome, which improved over time. There was no recurrence of lesion.

DISCUSSION

Paragangliomas are rare, slow-growing, neuroendocrine tumors that develop at autonomic ganglia. They originate from extra-adrenal paraganglia, which are derivatives of neural crest cells. In the head and neck region, these tumors most commonly occur at the carotid body (61.7%), jugular bulb (26.7%), and vagus nerve (9.3%).

We present two cases of cervical sympathetic chain paragangliomas bringing the total number of reported cases in the world literature to eleven (Table 1). The average age at presentation in the reviewed series is 22 years (range 8-46 years). Nine of the cases indicate whether there is presence of a pre-operative Horner’s syndrome, of which seven (79%) presented with this. Fifty percent (4 of 8) have functional symptoms or positive catecholamine laboratory testing. None of the reported cases mention details of the Doppler examination, but both of our patients had visible posterior pharyngeal submucosal fullness.

Imaging of the sympathetic cervical chain paragangliomas is characterized as being hypervascular on contrast CT and flow voids on T2 contrasted MRI. They tend to displace both the carotid and jugular vessels anterolaterally. The internal and external carotid arteries may be mildly splayed, depending on anterior extent of the mass. As this lesion is not bound by the carotid sheath, once large enough it may extend to the retropharyngeal space, as seen in our patients.

All cases in the literature were treated with resection of the mass with careful intraoperative monitoring of blood pressure, especially in cases with functional tumors.

CONCLUSION

Paragangliomas arising from the cervical sympathetic chain are exceptionally rare but must remain in the differential diagnosis of paragangliomas masses. Presentation of Horner’s syndrome and posterior pharyngeal fullness in addition to a lack of distinct imaging characteristics of schwannoma may warrant suspicion for paraganglioma of the cervical sympathetic chain. Treatment in all reported cases has been with surgical resection.

CASE 2

A 46 year old man presented with a four week history of swelling in the back of his throat and slowly enlarging right sided neck mass. He had onset of anisocoria and mild right-sided ptosis three years prior, and had been diagnosed with Horner’s syndrome at that time. He developed constant raspy hoarseness one year ago. He denied dyspnea, dysphagia, palpitations, hypertension, tachycardia, weight loss, and night sweats. He had no significant medical history, and no family history of paraganglioma.

Physical examination revealed anisocoria with constricted right pupil along with right eyelid ptosis (Figure 3). The right level II neck had a hard, immobile 5 cm mass. The right posterior and lateral oropharynx had a visible submucosal mass (Figure 4). On flexible nasopharyngoscopy, this mass extended a total of 5 cm along the right lateral pharyngeal wall.

Contrast CT showed an enhancing mass in the right parapharyngeal space measuring 5.2 x 3.1 cm that originated near the carotid bifurcation. The external and internal carotid artery (ECA and ICA) were mildly splayed at their bifurcation and superciliary the mass displaces both the vessels anterolaterally. Contrasted MRI reveals similar findings with mass on T2 Imaging (Figure 5).

The patient underwent complete resection of the mass via wide surgical exposure of the right neck. The mass was intimately adherent to the sympathetic trunk and did not involve the great vessels or vagus nerve (Figure 6). Selective neck dissection of levels II and III was performed.

Histologic analysis is consistent with paraganglioma (Figure 7). All lymph nodes were negative.

The patient recovered well with no neurological deficits besides continuation of the Horner’s syndrome. He did have first bite syndrome, which improved over time. There was no recurrence of the lesion.

BIBLIOGRAPHY