Turk Neurosurg 34(1):167-170, 2024

Case Report

DOI: 10.5137/1019-5149.JTN.40702-22.2

Received: 01.05.2022
Accepted: 27.07.2022

Published Online: 22.09.2023

Parapharyngeal Prestyloid Vagal Paraganglioma

Nuriye Guzin OZDEMIR[1], Hakan YILMAZ[2], Ibrahim Burak ATCI[1], Adil Can KARAOGLU[1], Okan TURK[1], Arzu Algun GEDIK[3], Ayhan KOCAK[1]

[1]University of Health Sciences, Istanbul Training and Research Hospital, Department of Neurosurgery, Istanbul, Türkiye
[2]University of Health Sciences, Izmir Bozyaka Training and Research Hospital, Department of Neurosurgery, Izmir, Türkiye
[3]University of Health Sciences, Istanbul Training and Research Hospital, Department of Pathology, Istanbul, Türkiye

Corresponding author: Hakan YILMAZ ✉ dr_hakanyilmaz@hotmail.com

ABSTRACT

Vagal paragangliomas (VPs) are rare tumors arising from paraganglionic tissue within the vagal nerve's perineurium. Usually, benign vascular tumors, VPs tend to invade the surrounding structures. Herein, we report the case of a VP presenting as a neck mass, which was evaluated as a glomus caroticum tumor preoperatively. A 65-year-old female complaining of a left-sided neck mass and intermittent hoarseness was assessed and operated on for possible glomus caroticum tumor. During the tumor excision, the vagal nerve was also involved, and hence, sacrificed. Histopathological examination revealed an encapsulated tumor associated with a nerve and ganglion and immunohistochemical staining tested positive for succinate dehydrogenase, confirming the diagnosis of VP. Postoperative residual hoarseness was corrected by vocal rehabilitation. While evaluating a retropharyngeal prestyloid neck mass, a VP should always be considered. Surgical excision involving vagal scarification, followed by vocal rehabilitation may be the appropriate treatment strategy.

KEYWORDS: Paraganglioma, Parapharyngeal, Prestyloid, Vagal

INTRODUCTION

Vagal paragangliomas (VPs) are rare neuroendocrine tumors arising from the neural crest-derived paraganglionic tissue surrounding the vagus nerve. Comprising less than 5% of all head and neck paragangliomas, patients with VP usually present with a neck mass pulsatile tinnitus or hoarseness (2). Approximately 2% of VPs can potentially become metabolically active to secrete catecholamines. They may also turn malignant and metastatic (3).

The primary treatment options include surgery and stereotactic radiosurgery, while somecases may be followed up without intervention (1). Herein, we present the case of a VP with a neck mass.

CASE PRESENTATION

Written informed consent was obtained from the patient for participation in the study and publication of data.

A 65-year-old female was admitted with an 8-week history of a neck mass and intermittent hoarseness. Dysphagia or velopharyngeal insufficiency was not reported. Physical examination revealed a 3.5 cm left-sided two-level mobile and nonpulsatile neck mass deep to the sternocleidomastoid muscle. Her laboratory results were normal and the 24-hour urinalysis was negative for catecholamines.

Further investigation with magnetic resonance imaging (MRI) showed a retromandibular-prestyloid mass (3.5×2.5 × 2.5 cm) on the left side. Computed tomography (CT) angiography and digital subtraction angiography demonstrated a neck mass within the left carotid sheath (Figures 1 and 2). The differential diagnoses for the condition included schwannoma, glomus tumor, neurofibroma, and VP.

The mass, suspected to be a glomus caroticum, was excised through an 8 cm submandibular incision over the sternocleidomastoid. A 3 × 2 cm highly vascular red mass

Nuriye Guzin OZDEMIR : 0000-0002-2702-4526
Hakan YILMAZ : 0000-0002-2180-1195
Ibrahim Burak ATCI : 0000-0002-0317-4159
Adil Can KARAOGLU : 0000-0001-9140-3005
Okan TURK : 0000-0002-0074-2835
Arzu Algun GEDIK : 0000-0002-9600-4289
Ayhan KOCAK : 0000-0002-3389-1941

was found between the carotid arteries anterolaterally and the internal jugular vein posteromedially. The tumor was displacing the left internal carotid artery forward and the left internal jugular vein backward. The dissection was made along the internal and external carotid arteries and the internal jugular vein, preserving the spinal accessory and hypoglossal nerves. The vagal nerve was involved within the mass and removed with the mass after ligation (Figure 3). During this phase, the lesion was assumed as a vagal schwannoma.

The histopathological report of the resected specimen described an encapsulated tumor (3.5×2.5×2.5 cm) associated with a nerve and ganglion. The central 5% of the tumor showed necrosis. Immunohistochemical staining for succinatedehydrogenase (SDH) was positive, and a diagnosis of VP was made (Figure 4).

After the surgery, she had no dysphagia and tolerated her regular diet; however, she was referred to the Ear, Nose, and Throat (ENT) outpatient clinic for evaluation of hoarseness for vocal rehabilitation.

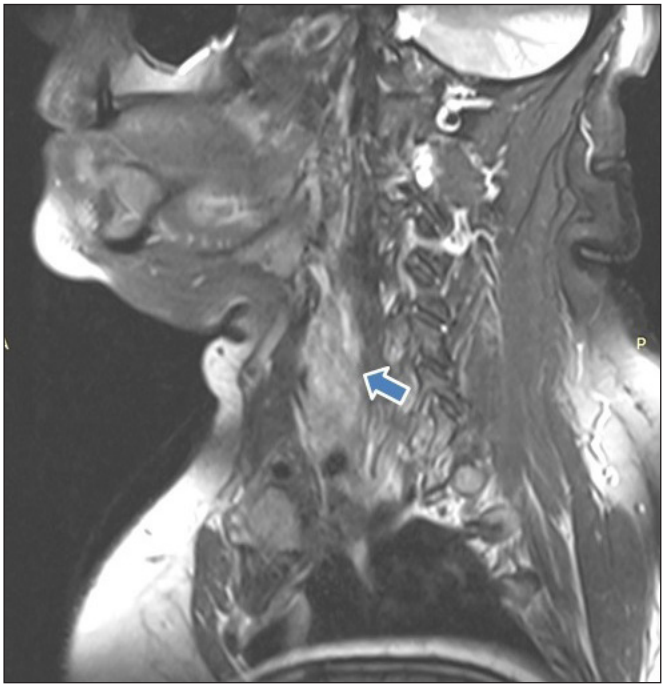

Figure 1: T-2 weighted sagittal magnetic resonance imaging of the neck showing left prestyloid mass (blue arrow).

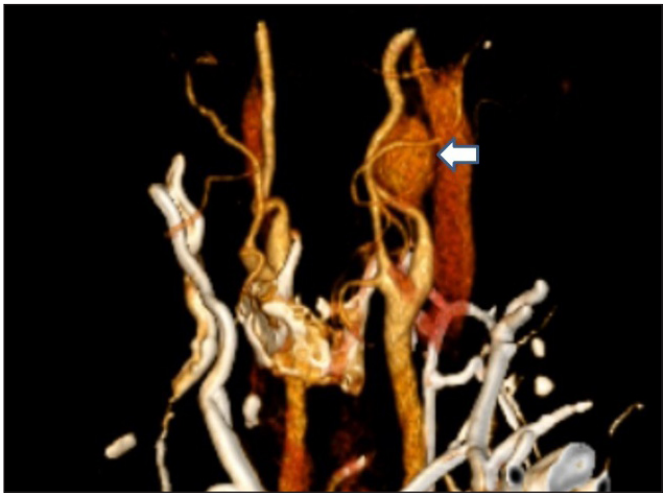

Figure 2: Angiographic view of vagal mass (white arrow).

DISCUSSION

Paraganglia are small groups of neuroendocrine cells stemming from the autonomic nervous system ganglia (7). Arising from the paraganglia, paragangliomas are usually benign and most commonly located in the adrenal medulla; only 3% are located in the head and neck. They may arise in the carotid body, vagal nerve, middle ear, and larynx. More than half of head and neck paragangliomas are sporadic without a genetic cause. VPs are usually rare, slow-growing benign vascular tumors; however, they tend to invade the surrounding structures (9) and proximity to the lower cranial nerves may risk the surgery. Additionally, bilaterality and multicentricity may be commonly observed. Although head and neck paragangliomas are generally non-functional, 4% were reported to be functional (9). The differential diagnoses for VPs include schwannoma, meningioma, carotid paraganglioma, and jugulo-tympanic paraganglioma (2).

Four genetic paraganglioma syndromes have been described in the literature with an autosomal dominant transmission. Three of these syndromes have germline mutations in the

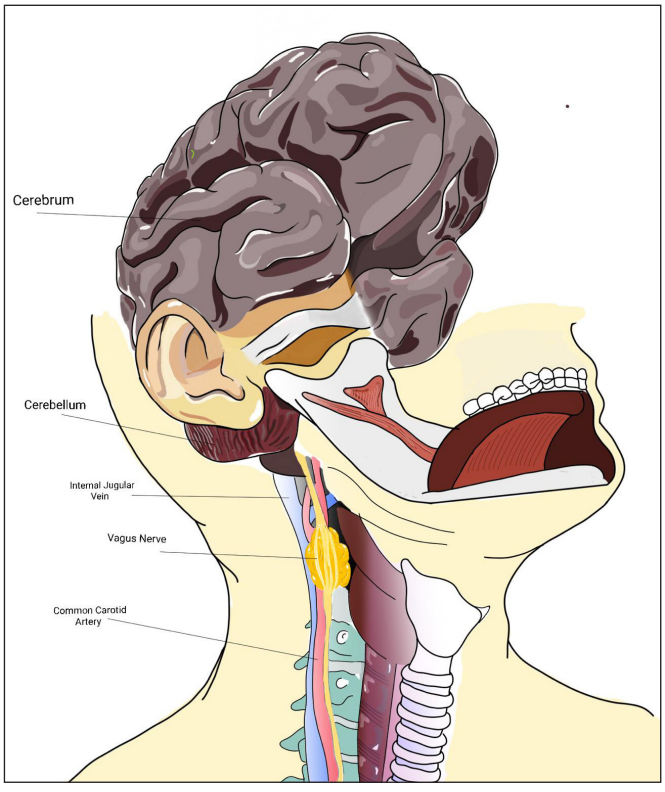

Figure 3: Demonstration of surgical anatomy.

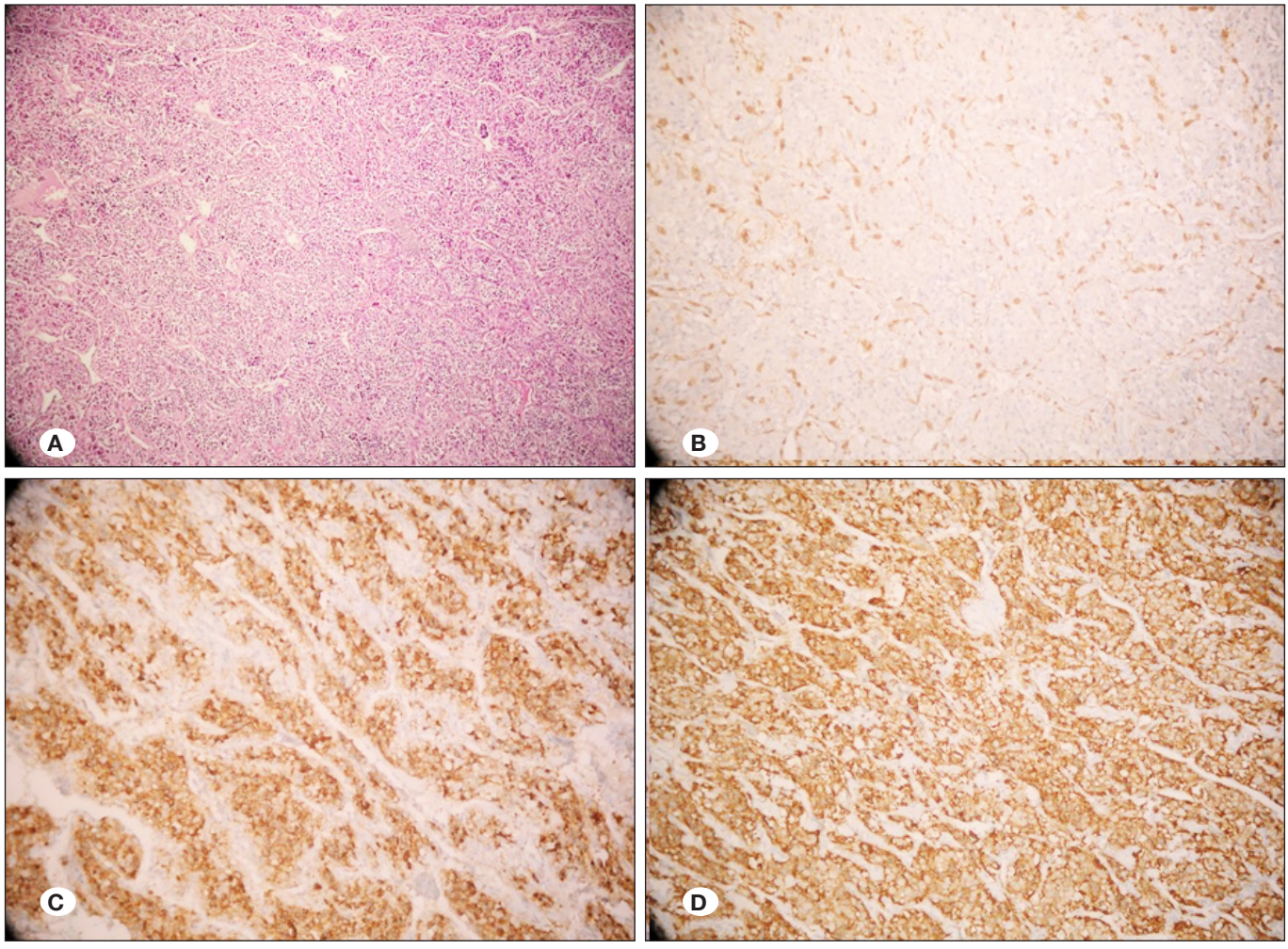

Figure 4: A) **Hematoxylin and Eosin (H&E)(x10):** The tumor cells form nests known as zellballen within a fibrovascular stroma. The sustentacular cells are not identified on H&E staining but are prominent with S100 staining. **B) S100:** Paragangliomas may show diffuse weak variable cytoplasmic staining in the chief cells but the sustentacular cells are usually strongly positive. **C) Chromogranin A:** Paragangliomas have diffuse strong cytoplasmic positivity. **D) Synaptophysin:** Paragangliomas have diffuse strong cytoplasmic positivity (H&E, ×200).

gene complex encoding SDH in the SDH-D, SDH-C, or SDH-B subunits (8). These SDH mutations cause hypoxia leading to carotid paraganglial hypertrophy.

CT-angiography and MRI help localize the tumor and its extension. MRI is also useful for differentiating from the other parapharyngeal neurogenic and salivary gland neoplasms, besides being used as a screening tool for other family members. Contrast-enhanced 3D-MR angiography is reported as a more specific and non-invasive diagnostic technique for screening and follow-up of VPs (4,5). Volumetric analysis of these tumors has also been studied; however, anatomical changes in relation to surrounding structures were deemed as clinically more relevant than volumetric changes (2).

Surgical excision is the treatment of choice for VPs; however, since the vagus nerve is mostly involved in the tumor mass, resection may result in speech and swallowing deficits. VPs<3 cm in diameter may be treated with stereotactic radiosurgery, whereas observation is recommended in asymptomatic older patients. Neuropathy progression has been reported in a third of the cases (2).

During VP resection surgery, carotid artery exposure is necessary to avoid vascular complications. In the case of cranial base involvement, a lateral transtemporal or combined transcervical and transmandibular approach may be used. The styloid process may be resected in high cervical tumors. Additionally, mastoidectomy and sigmoid sinus ligation to allow jugular bulb mobilization may be performed for cranially and foraminally extended tumors (3). Since paragangliomas are highly vascular, preoperative embolization may help during the resection (6).

While surgical treatment is the best option, elderly patients may be managed conservatively owing to the difficulty in treating lower cranial nerve complications in this group. External beam or stereotactic treatment are good choices

for bilaterally located vagal tumors (3). In our patient, surgical excision was chosen due to the neckmass, and the symptoms were relieved with total excision; only minor hoarseness persisted after the operation.

Postoperatively, swallowing and speech rehabilitation is usually needed, as adopted in ourpatient. In the early postoperative days, she was monitored closely for swallowing difficulties and swallowing without aspiration. Primary medialization laryngoplasty is necessary for speech disturbance; however, vocal rehabilitation by the ENT surgeon sufficed for ourcase. In case of palatal paralysis due to involvement of the palatal branch of the vagus nerve, compensation is usually observed within 4–6 months with the help of swallowing therapy. In decompensated patients, palatal adhesion may be necessary (2).

Other complications include severe pain in the parotid region or face, and gastrointestinal and baroreceptor dysfunction; Horner syndrome may be also observed (3). Parotid involvement may result from damaged cervical sympathetics. Baroreceptor dysfunction is related to carotid sinus damage, while gastrointestinal dysfunction may be due to the visceral, sensory, motor, and parasympathetic innervations impairment in vagal scarification (3).

Therefore, surgical excision of VPs must be considered in clinically indicated cases; observation and radiotherapy are other options for the elderly and previously operated complicated cases (9).

CONCLUSION

While a retropharyngeal prestyloid neck mass may be evaluated as a glomus tumor, VP should always be considered as the differential diagnosis. In the case of the latter, surgical excision is the best treatment despite requiring vagal sacrification. Such a patient maybe followed up with an ENT surgeon for vocal rehabilitation.

AUTHORSHIP CONTRIBUTION

Study conception and design: NGO, HY, IBA, ACK, OK, AAG, AK
Data collection: NGO, HY
Analysis and interpretation of results: NGO, HY, IBA, ACK, OK, AAG, AK
Draft manuscript preparation: NGO, HY, IBA, ACK
Critical revision of the article: NGO, HY, IBA, ACK, OK, AAG, AK
Other (study supervision, fundings, materials, etc...): NGO,HY
All authors (NGO, HY, IBA, ACK, OK, AAG, AK) reviewed the results and approved the final version of the manuscript.

REFERENCES

1. Basaran B, Polat B, Unsaler S, Ulusan M, Aslan I, Hafiz G: Parapharyngeal space tumours: The efficiency of a transcervical approach without mandibulotomy through review of 44 cases. Acta Otorhinolaryngol Ital 34:310-316, 2014
2. Bhatt S, Verma P, Meena N, Tandon A, Tanveer N, Bansal D: Glomus vagale tumour, can it be diagnosed only on sonography? J Ultrasound 20:73-79, 2017
3. Caldarelli C, Iacconi C, DellaGiovampaola C, Iacconi P, Beatrice F: Vagal paragangliomas: Two case reports. Acta Otorhinolaryngol Ital 27:139-143, 2007
4. Corssmit EP, Romijn JA: Clinical management of paragangliomas. Eur J Endocrinol 171:R231-243, 2014
5. Heyes R, Taki N, O'Leary MA: Vagal paraganglioma presenting as a neck mass associated with cough on palpation. Case Rep Otolaryngol 2017:7603814, 2017
6. Hornbeek H, Iyer NG, Carlson DL, Stambuk HE, Ryder M, Ganly I: Functional vagal paraganglioma: A case report illustrating diagnosis and management. Skull Base 20:491-496, 2010
7. Hu K, Persky MS: Treatment of head and neck paragangliomas. Cancer Control 23:228-241, 2016
8. Singh D, Krishna PR: Paraganglioma of the vagus nerve mimicking as a carotid body tumor. J Vasc Surg 46:144, 2007
9. Zanoletti E, Mazzoni A: Vagal paraganglioma. Skull Base 16:161-167, 2006