Hypoglossal Nerve Palsy As an Initial Presentation of Glomus Jugulare Tumor in Patient with Breast Cancer: A Case Report and Literature Review

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Abstract

Background: Glomus jugulare tumor is a rare, slow-growing, hyper-vascular paraganglioma that originates from the neural crest derivatives in the wall of the jugular bulb. The most common clinical manifestations of glomus jugulare are pulsatile tinnitus, conductive hearing loss, and hoarseness due to its vascularity and invasion of surrounding structures. Isolated hypoglossal nerve palsy as a presenting feature of the glomus jugulare is very rare. Case Report: We report a 61-year-old woman with a past medical history of breast cancer and diabetic mellitus presenting with progressive difficulty handling food in her mouth and tongue atrophy. Investigations showed skull base lesion and solitary pulmonary nodule. Further work-up led to glomus jugulare and benign solitary pulmonary fibrous tumor diagnosis, although the first impression was metastatic involvement of the jugular foramen. Endovascular embolization of the glomus jugulare was performed, but the patient refused any open surgery due to co-morbidities and the risk of operation. She had no new symptoms at the one-year follow-up, and the size of the lesion became more minor on the follow-up imaging relative to the baseline. Conclusion: Glomus jugulare tumors should be considered and surveyed in the diagnostic work-up of patients with hypoglossal nerve palsy.

Keywords: Glomus Jugulare Tumor; Paraganglioma; Jugular Foramen; Hypoglossal Nerve Palsy

Introduction

Jugular foramen (JF) tumor is a rare, deeply seated, cranial base lesion that divides into primary and secondary according to its locations and extension from JF into the surrounding structures or vice versa [1]. The four most common neoplasms of the JF are paraganglioma, schwannoma, meningioma, and metastasis [2]. Paragangliomas have different nomenclature, including glomus jugulare, carotid body tumor, glomus vagal, and glomus tympanicum based on their locations in the head and neck (JF, carotid...
body, along the vagal nerve, and middle ear, respectively) [1]. Glomus jugulare tumors occur preferentially in the sixth decade of life with a female to male ratio of 5:1 [3]. The most common clinical presentations of glomus jugulare are pulsatile tinnitus and conductive hearing loss [4]. Twelfth cranial nerve palsy as an initial presentation of glomus jugulare is very rare, and thus far, only a few cases have been reported [3,5-7]. In this unique case report, we describe a patient with a past medical history (PMH) of breast cancer who developed glossal atrophy, which was proved to be caused by a glomus jugulare tumor. The patient had different types of tumors concurrently.

**Case Presentation**

A 61-year-old woman was admitted to the emergency department of Shariati hospital, affiliated with Tehran University of Medical Sciences, Tehran, Iran. She complained of progressive difficulty in eating performance two months before the admission. She had no history of tinnitus, hearing loss, and/or hoarseness. Her PMH was significant for diabetes mellitus and breast cancer. She underwent a modified radical mastectomy and chemotherapy about eight years ago, and her cancer was in remission. Drug history was consistent with metformin 500 mg twice daily. On arrival at the hospital, her vital signs were normal, and she was oriented to time, place, and person. Neurological examination revealed atrophy of the left side of the tongue with deviation to the left on protrusion consistent with left-sided hypoglossal nerves palsy. Besides, mildly reduced gag reflex on the left side was noted. The rest of the neurological examination was unremarkable. Due to her clinical symptoms,

![Figure 1. A and B: Baseline brain MRI on T1W and T2W, respectively, reveal a 20×12×10 mm mass with signal flow voids and salt and pepper appearances in the left jugular foramen (thick white arrows). C: Brain MRI on T1W with contrast demonstrates avid enhancing lesion in the left jugular foramen (thick white arrows). D: Super selective ascending pharyngeal artery angiogram shows a sizeable vascular mass with intense tumor blush (thick white arrow) and early draining veins (thin white arrows).](image-url)
and neurological examination findings, brain magnetic resonance imaging (MRI) was performed, which showed a 20×12×10mm extra-axial mass with heterogeneous signal intensity and intense contrast enhancement within the left JF (Figure-1A-C). Considering patient’s PMH and the skull base lesion in brain MRI, cerebrospinal fluid (CSF) analysis, malignancy work-ups, including spiral chest computed tomography (CT) scan, abdominopelvic CT scan, bone scintigraphy, breast sonography/mammography, and tumor markers as well as vasculitis markers, and angiotensin-converting enzyme (ACE) was requested. CSF opening pressure was 15 cm of H2O, and analysis showed normal white blood cell (WBC) count, protein, glucose, and lactate dehydrogenase (LDH) with negative cytology. The complete blood count (CBC), LDH, erythrocyte sedimentation rate (ESR), vasculitis markers comprising anti-nuclear antibody (ANA), anti-double strand DNA (anti-dsDNA), anti-Ro, anti-La, antineutrophil cytoplasmic antibodies (ANCA), and ACE level were normal. A spiral chest CT scan confirmed the presence of a 20×30mm nodule in the apical segment of the right lower lobe (Figure-2A). Abdominopelvic CT scan, skeletal scintigraphy, breast sonography/mammography, and tumor markers (carcinoembryonic antigen [CEA], cancer antigen-125 [CA-125], CA19-9, and alpha-fetoprotein [AFP]) were normal. As a result of the patient’s PMH and imaging findings, metastasis was considered as the initial diagnosis. Pathologic examination of
CT-guided biopsy from pulmonary nodule demonstrated a benign solitary pulmonary fibrous tumor (Figure-2B), which was reviewed and confirmed in another center. The MR images were double-checked, and the diagnosis of glomus jugulare was considered due to the salt-and-pepper appearance and flow voids on T1-weighted (T1W) and T2-weighted images (T2W). Consequently, angiography was performed that affirmed the diagnosis of glomus jugulare (Figure-1D), and embolization of the tumor was made during the procedure.

Because of her co-morbidities and high risk of surgery, the patient preferred to be followed by clinical examination and MR imaging. At the follow-up visit after one year, her symptoms did not change, but the tumor became shrinkage and smaller in size 16×10×8mm (Figure-3A and B), which we interpreted as a result of embolization. This study was approved by the ethics committee of Shiraz University of Medical Sciences (approval number:IR. sums.med.rec.1400.216). Informed consent was obtained from the patient to publish her anonymous data in this case report.

Discussion

As a result of broad-spectrum differential diagnosis of skull base pathologies, a preoperative diagnosis is essential as there are significant differences in the treatment and surgical strategies for each of these lesions. In the present case, considering the patient’s PMH and MRI findings, the most probable differential diagnosis is metastasis followed by primary JF tumors and less likely vasculitis and inflammatory disorders such as sarcoidosis. Given negative vasculitis markers, ACE, normal CSF analysis, and skull base lesion imaging pattern, we ruled out the likelihood of vasculitis and inflammatory disorders. Besides, the other differential diagnosis is metastasis, and the most common tumors that involve the skull base are breast, lung, prostate, and malignant lymphoma [8]. Therefore, extensive malignancy work-ups were performed, which showed only a solitary pulmonary fibrous tumor. Due to the result of malignancy work-ups, MRI characteristics of the lesion, and angiography findings, the possibility of metastasis was ruled out.

Furthermore, primary JF tumors, including lower cranial nerves schwannomas, meningioma, and glomus jugulare, are the other differential diagnosis. Schwannomas and meningiomas appear as T1W hypointense and T2W hyperintense lesions on MRI with mild to moderate vascularity on cerebral angiography and do not invade adjacent bony structures [1]. The imaging characteristics of the glomus jugulare tumor are salt and pepper appearance and signal flow voids on T1W and T2W sequences, with intense enhancement after contrast administration [1]. Glomus jugulare appears as a hyper-vascular mass with enlarged feeding arteries, intense tumor blush, and early draining veins on angiography [9], likely to our presenting case. Due to the imaging findings and the work-up results, a diagnosis of glomus jugulare was made. Glomus jugulare tumors originate from the glomus body in the adventitia at the dome of the jugular bulb [10].

Although usually histologically benign, they are characterized by diverse biological behavior and have a tendency for focal invasion [10]. The most common initial symptoms are pulsatile tinnitus (84.3%), followed by conductive hearing loss (75.9%) and hoarseness (34.9%) [4]. Hypoglossal nerve paralysis commonly occurs nine years after the initial presentation of glomus jugulare, and its occurrence as an initial manifestation of glomus jugulare in isolation or association with other cranial nerve palsies is infrequent. Thus far, only a few cases have been reported in the literature [3,5-7]. Only two of these reported cases showed isolated hypoglossal nerve palsy similar to our reported patient. Most of the previously reported [3,6,7] cases with hypoglossal nerve palsy in association with glomus jugulare are female consistent with our presented case. Although widespread investigations have been done on the treatment modalities of glomus jugulare tumors, sometimes the most favorable route of management remains controversial [10]. Preoperative embolization seems safe and
increases the chance of complete resection of the head and neck hyper-vascular tumors and minimizes blood loss during surgery; high-risk patients for surgery may benefit from palliative embolization [11]. In individuals with significant co-morbidities or large tumors, external beam radiotherapy and/or gamma knife stereotactic radiosurgery is another treatment strategy that can be used as monotherapy or adjunctive therapy with acceptable risk and effectiveness [12,13]. Glomus jugulare should be considered in the differential diagnosis of any patient with isolated hypoglossal nerve palsy, and neurologists should be familiar with the imaging characteristics of glomus tumors.

**Conclusion**

Our case report illustrates the importance of proper knowledge about clinical and radiological manifestations of glomus jugulare tumors. Glomus jugulare should be considered and scrutinized in the diagnostic work-up of patients who develop hypoglossal nerve palsy, even though the PMH is consistent with malignancy in other parts of the body.

**Conflict of Interest**

None.

**References**