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Mixed Cranial Nerve Schwannoma: A Rare entity Recognition and Key Imaging Features

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Abstract

Introduction: Schwannomas of mixed cranial nerves, often referred to as jugular foramen schwannomas, are rare tumors. Their clinical presentation is usually nonspecific, which can easily be confused with other lesions. In particular, jugular paragangliomas represent the main differential diagnosis. The aim of this study is to clarify the diagnostic approach for these schwannomas and to establish criteria for differentiating them from paragangliomas.

Case report: A 82 years old female patient, was referred to our department for progressive swallowing difficulties, alteration of taste sensation and mixed dysphagia and dysphonia, Without dyspnea or any associated signs. The clinical examination revealed right-sided spinal accessory nerve paresis and deviation of the soft palate to the left and a right vocal cord paralysis, these findings were consistent with a lesion involving the mixed cranial nerves. Brain MRI revealed an enlarged right jugular foramen with smooth osseous remodeling, without bone erosion or destruction. occupied by a well-defined tissue mass with marked enhancement after contrast administration. The patient was discussed at a staff meeting where a diagnosis of schwannoma involving the mixed cranial nerves was established. And Gamma Knife radiosurgery was recommended as the therapeutic approach.

Conclusion: The main points emphasize that imaging plays a crucial role in the diagnosis of jugular foramen schwannomas and can alone allow differential diagnosis from paragangliomas. Schwannomas typically appear as homogeneous soft tissue masses with regular borders, centered on the jugular foramen, and may extend to the cerebellopontine angle. They show no signs of bone destruction and lack the characteristic "salt-and-pepper" appearance seen in paragangliomas.

Keywords: Jugular foramen schwannoma; Mixed cranial nerve tumor; Magnetic Resonance Imaging (MRI); Differential diagnosis; Paraganglioma

Introduction

Schwannomas of mixed cranial nerves are rare pathological entities that may develop in the cervical region, the jugular foramen, or the hypoglossal canal. The parapharyngeal space represents the most frequent location, accounting for 31% of cases, followed by the cervical region (23%), whereas the skull base is involved in only 19% of cases. These tumors may present with a constellation of cranial neuropathies, including dysphagia, phonatory disorders, and shoulder weakness, reflecting the involvement of multiple nerves within an anatomically complex region. The differential diagnosis includes jugular paragangliomas, which are highly vascularized and exhibit distinctive radiological features. A rigorous correlation between clinical and radiological findings is essential to establish an accurate diagnosis and to guide therapeutic decision-making, particularly when considering minimally invasive treatments such as stereotactic radiosurgery using the Gamma Knife.

Case Presentation

A 82 years old female patient, with no particular pathological antecedents, was referred to our department for progressive swallowing difficulties, alteration of taste sensation and mixed dysphagia and dysphonia, evolving over the past two months. Without dyspnea or any associated signs.

The clinical examination revealed right-sided spinal accessory nerve paresis and deviation of the soft palate to the left, without facial palsy, vertigo, parapharyngeal or cervical mass, or lymphadenopathy.

Furthermore, nasofibrolaryngoscopic evaluation demonstrated right vocal cord paralysis. The facial and cervical examination was otherwise unremarkable. These findings were consistent with a lesion involving the mixed cranial nerves **(Figure 1)**.

A Brain MRI was performed and revealed an enlarged right jugular foramen with smooth osseous remodeling, without bone erosion or destruction. occupied by a well-defined tissue mass showing spontaneous T1 hyperintensity and T2 hypointensity, with marked enhancement after contrast administration. The lesion measured 23 × 20 mm and

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extended into the right cerebellopontine angle, compressing the ipsilateral acoustic–facial bundle **(Figure 2)**.



Figure 1: Image showing the patient's neck with no evidence of a cervical mass or cervical lymphadenopathy.

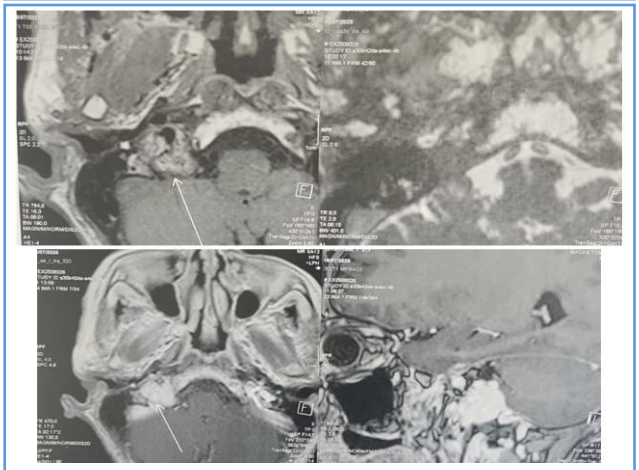


Figure 2: Axial / sagittal sections of brain MRI showing the right shwannoma of the mixed cranial nerves centered on the right jugular foramen.

The patient was subsequently presented at a multidisciplinary team meeting, where a diagnosis of schwannoma involving the mixed cranial nerves was established. Accordingly, the team recommended Gamma Knife radiosurgery as the therapeutic approach. based on a set of arguments, particularly the complexity of surgical management in an elderly patient over 80 years of age, with a large mass in close intimate contact with the internal carotid artery.

The diagnosis of schwannoma was favored based on the integration of clinical and radiological findings:

Radiologically, the lesion was well-circumscribed, exhibited spontaneous T1 hyperintensity and T2 hypo intensity, and showed homogeneous contrast enhancement without any bone destruction.

Importantly, the mass was extended into the right cerebellopontine angle, a feature more consistent with a schwannoma than a paraganglioma.

Clinically, the selective involvement of the glossopharyngeal, vagus, and spinal accessory nerves including right vocal cord paralysis and deviation of the soft palate further supported the diagnosis of a mixed cranial nerve schwannoma rather than a paraganglioma.

The patient showed a favorable clinical course following Gamma Knife radiosurgery, with gradual regression of neurological deficits. Swallowing difficulties and voice changes improved, and overall clinical status showed significant improvement during follow-up.

Discussion

Glossopharyngeal schwannomas are rare tumors, most commonly arising from the intracranial region, particularly the cerebellopontine angle. The few reported extracranial schwannomas generally result from secondary extension of intracranial lesions [1]. Schwannomas of cranial nerves IX to XI are often grouped under the term jugular foramen schwannomas, and only intraoperatively are they specifically identified. These tumors are extremely rare, especially in the absence of neurofibromatosis [1,2].

Mixed cranial nerve schwannomas frequently lack specific symptoms or pathognomonic clinical signs, making diagnosis challenging [3]. Their clinical presentation is variable, and preoperative suspicion typically relies on the combination of clinical manifestations and radiological findings. Symptoms such as hearing loss, dysphagia, ageusia, dysphonia, pain, or compression signs generally appear only when the tumor reaches a significant size. These schwannomas may occur sporadically or in the context of familial tumor syndromes such as NF2 or schwannomatosis, with malignant transformation being exceedingly rare [4]. The differential diagnosis includes paragangliomas, neurogenic tumors, carotid body tumors, and neurofibromas [1]. Radiologically, smooth bony remodeling, relatively homogeneous signal, and intense contrast enhancement [5] generally characterize schwannomas. In contrast, paragangliomas typically present with permeative bone erosion, intense enhancement, and a "salt-and-pepper" appearance on T1-weighted sequences due to subacute hemorrhage [5]. However, some small lesions may lack these typical features, complicating diagnosis. Despite the precision of neuroimaging, imaging alone is often insufficient for a definitive diagnosis of a cranial nerve IX schwannoma,

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and intraoperative identification frequently remains necessary. Furthermore, advanced dynamic imaging techniques (*e.g.,* GRASP MRI) can aid in differentiating schwannomas from paragangliomas based on vascularization and enhancement parameters [6]. For treatment, the distinction between schwannomas and paragangliomas is crucial, as it may alter the therapeutic strategy [6,7]. Surgical excision remains the treatment of choice for glossopharyngeal and vagal nerve schwannomas, with preservation of these nerves whenever feasible. For residual tumors or in cases where surgery is considered high-risk or the patient is deemed inoperable, stereotactic radiosurgery, particularly Gamma Knife, represents an effective alternative [4].

Conclusion

Schwannomas should always be considered in the differential diagnosis of lesions at the jugular foramen and not overlooked in favor of paragangliomas. Their nonspecific clinical presentation makes imaging interpretation crucial. MRI allows detailed evaluation of tumor characteristics, helping to distinguish schwannomas from other highly vascular lesions. Multidisciplinary discussion of each case ensures that the diagnosis is not missed and that an optimal management plan is established. Early recognition and careful planning are essential for implementing an appropriate therapeutic strategy and improving patient outcomes.

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