# Lower Cranial Nerve Schwannomas

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## Abstract

Lower cranial nerve (LCN) schwannomas are rare, benign peripheral nerve sheath tumours arising from cranial nerves IX–XII. Though uncommon, they pose significant clinical challenges due to their complex anatomical location and potential for neurological deficits. Jugular foramen schwannomas (JFS) and hypoglossal schwannomas (HS) are the most prevalent subtypes. Clinical presentation varies by nerve involvement, ranging from hoarseness and dysphagia to tongue atrophy and paroxysmal cough. Diagnosis relies heavily on MRI and CT imaging, with differentiation from paragangliomas being critical. Classification systems guide surgical planning, with gross total resection (GTR) being the goal, though subtotal resection (STR) is preferred in cases with dense neural adherence to preserve function. Gamma knife radiosurgery (GKRS) offers an effective alternative for small or residual lesions. Treatment must balance tumor control with preservation of neurological function, requiring a multidisciplinary and individualized approach.

**Keywords:** Schwannoma, Benign Neoplasms, Cranial nerve neoplasms

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## Introduction

Schwannomas are benign tumours arising from Schwann cells of peripheral nerves.[1] While vestibular schwannomas are the most common, sporadic non-vestibular cranial nerve schwannomas (NVCNS) account for only 5–10% of cases, with lower cranial nerve schwannomas (LCNS) being exceptionally rare.[2,3] LCNS may involve cranial nerves IX (glossopharyngeal), X (vagus), XI (accessory), or XII (hypoglossal). The majority arise in the jugular foramen (jugular foramen schwannomas, JFS), while others rarely originate in the hypoglossal canal (HS) or cervical region.[4] Clinically, LCNS often present as slow-growing, asymptomatic neck masses but may cause compressive symptoms or nerve deficits, including dysphonia, dysphagia/odynophagia, nasal obstruction/epistaxis, shoulder/upper extremity pain, or even respiratory compromise.[4] Malignant transformation is rare, and surgical resection is typically curative with a low recurrence rate.[2] However, due to the critical functions of the lower cranial nerves, postoperative complications are common. Although most cases are solitary, multifocal schwannomas should raise suspicion for neurofibromatosis type 1 (NF1), type 2 (NF2), or schwannomatosis, necessitating genetic evaluation.[5]

## Review of Evidence:

LCN schwannomas present as a therapeutic challenge due to the critical anatomical location, the complex surrounding neurovasculature and the potential for significant neurological morbidity.[6,7] Clinical presentation varies depending on the nerve of origin and tumour's anatomical spread.[8] The most common preoperative symptoms as reported by Hoffman et al., in their systematic review were hearing loss (52.2%), hoarseness (47.3%), dysphagia (39%), gait impairment (26.2%), headache (24%), facial weakness (12.5%), and facial numbness (9.5%).[9] Hypoglossal schwannomas are characterised by unilateral tongue atrophy and fasciculations while vagal schwannomas present with the hallmark sign of paroxysmal cough upon palpation of the tumour.[10]

Radiologic assessment using magnetic resonance imaging (MRI) and fine sliced computed tomography (CT) scans is a cornerstone of diagnosing these tumours. CT is important in evaluating bony erosions and skull base foraminal remodelling.[7] On the other hand, MRI provides an in depth soft tissue characterisation and tumour extension. Hypointense or isotense signals on T1-weighted MRI and hyperintense signals on T2 with homogenous enhancement on gadolinium administration is diagnostic of these tumours.[2,10] Paragangliomas are a major radiological differential. In comparison, Schwannomas differ however by their lack of flow voids and lytic bone destruction. Exrtracranial vagal schwannomas are also found in the parapharyngeal spaces.[4,5] These tumours tend to show similarity to carotid body and glomus vagale tumours. However, on MRI these tumours are shown to displace the carotid artery anteromedially, distinguishing from the paragangliomas.[11]

The pattern of tumour expansion can assist in distinguishing between these lesions. Paragangliomas

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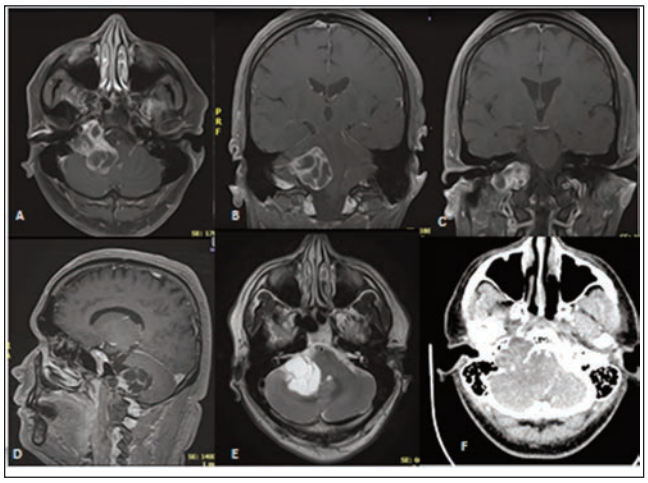

**Figure:** A-F: The images are of a 46-year-old gentleman who presented with progressively worsening headache and recent onset vomiting. Fig A shows a T1 post contrast axial image showing a right cerebellopontine angle lesion with multiple enhancing cystic components and extension into the right jugular foramen and expansion thereof. Fig B and C show similar findings on coronal post contrast images. Fig D is sagittal post contrast image showing involvement of right jugular foramen. Fig E shows a T2 axial image in which a normal VII and VIII cranial nerve complex can be seen ruling out vestibular schwannoma. Fig F shows a CT brain post contrast axial image involving erosion of right petrous bone, mastoid and clivus with expansion of jugular foramen. Radiological diagnosis was schwannoma of lower cranial nerves. Intra-operative findings validated the radiological images and histopathology confirmed Schwannoma.

usually grow in a superolateral direction from the jugular foramen toward the middle ear. In contrast, schwannomas tend to extend superomedially toward the brainstem, aligning with the trajectory of the lower cranial nerves. On bone CT imaging, schwannomas are typically characterized by sharply defined margins with peripheral sclerosis, while paragangliomas more commonly exhibit irregular, erosive bone destruction.[4]

Various classification system aid in surgical planning. Kaye et al., and Samii et al., categorised JFS into types A to D based on tumour spread.[4] Type A is purely intracranial, type B is intraosseous, type C extracranial, and type D involving all three levels. This system was further refined by Samii et al., with subtypes B1 to B3, depending on the intraosseous and extraforaminal spread.[4] A similar anatomical classification was developed by Nonaka et al for hypoglossal schwannoma.[8] These systems are extremely crucial in delineating different approaches when planning the surgical strategy and predicting outcomes.[8]

The primary management modality of LCN schwannomas is surgical resection. The aim of surgery is to achieve gross total resection (GTR) while preserving neurological function. The approach depends on the tumour location and extension.[2] Retrosigmoid approach is commonly used for intracranial tumours, classified as type A by the aforementioned classification systems, while infralabyrinthine or transcondylar approaches are necessary for types B, C and D which present with intraosseous and extending dumbbell shaped lesions.[2,4,12] Endoscopic assistance is increasingly becoming a part of the modality for better visualisation of the deep corridors and reducing intra-operative complications. A more complicated management arises in cases of multiple schwannomas particularly involving cranial nerve X and XII.(5) Due to predisposing NF1 or NF2, multiple schwannomas are harboured in these regions resulting in increased risk of multiple nerve injuries. Verma et al proposed use of nerve sparing techniques by utilising microscopic magnification.[5] Subtotal resection (STR) with intraoperative mapping is essential for decreasing morbidity. Robotic approaches have been defined in cases of extracranial vagal schwannomas which are best managed surgically when patient is symptomatic.[5,11]

Hoffman et al., in their systematic review reported 81% pooled GTR rates for JFS.[9] However, lower cranial nerve deficits still remain a major concern. Among the most debilitating complications is damage to cranial nerve IX and X resulting in postoperative dysphagia and hoarseness.[9] Sometimes tracheostomy and feeding tubes may also be needed for temporary improvement of acute exacerbations.[7] In comparison to new defects, patients with preoperative deficits tend to adapt over time. On the other hand, in HS, due to the complicated course of the cranial nerve XII, surgery is far more complex. In case of dense adherence of the tumour to the nerve, STR is the treatment of choice.[5] Hoffman et al., and Oyama et al., have documented the use of sural nerve grafts for hypoglossal nerve repair and also achieving partial recovery. Similarly to JSF, endoscopic approaches are have been explored for dumbbell shaped lesion however they come with risks of CSF leak and vascular injury.[3,9]

Radiosurgery, in particular gamma knife radiosurgery (GKRS), has proven to be an effective non invasive treatment option for LCN schwannomas. Hofman et al., reported a review of 246 patients treated with GKRS and reported successful tumour control in 98% of the cases.[9] Neurological deficits were seen in 34% of the cases post interventions of which most were transient. According to the reports, preferable outcomes can be expected with radiosurgery for small (<6 $cm^3$), non extending tumours or residual lesion post STR.[2,4] It may also present as an alternative in the elderly population that do not qualify for open surgery. Recent data as presented by Sedney et al. suggest that near total resections with preserved functions often do not require upfront radiosurgery, with low recurrence rates. These findings support a shift towards less

radical strategies in cases with high adherence to vital nerves.[4]

Histologically, schwannomas exhibit Antoni A (hypercellular) and Antoni B (hypocellular) areas. Verocay bodies, a hallmark of the Antoni A pattern, are characterized by palisading arrangements of tumour cell nuclei flanking a central acellular zone, creating the appearance of parallel rows separated by clear spaces.[5] S100 immunopositivity confirms Schwann cell origin. Except for NF2 patients, malignant transformations are extremely rare and should only be considered in rapidly enlarging lesions like that of NF2. Radical excision with adjuvant therapy is the best treatment choice in these patients.[5]

## Conclusion:

LCN Schwannomas although rare but they are clinically significant tumours requiring individualised multidisciplinary management. Advances in surgical approaches alongwith intraoperative mapping have improved outcomes and decreased postoperative nerve deficits as a common complication of surgery. In addition, radiosurgery also provides a viable alternative and as an adjunct for complete removal of the lesion. The decision making process must balance the goals of tumour control and functional preservation. The differing severity and complications require a cautious and personalised approach to each case.

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