

# Navigating the Diagnostic Challenges of Cervical Plexus Schwannoma: A Rare Entity

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## Keywords

Schwannoma, Cervical Schwannoma, Posterior neck mass, Nerve sheath tumours, Cervical plexus

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## INTRODUCTION

Schwannoma is a mesenchymatous type of tumour that develops from the Schwann cells. Schwannomas are part of the peripheral nerve sheath tumours. They are slow-growing and can originate from the cranial, peripheral, or autonomic nerves. Cervical schwannoma, in general, has a growth rate of 5.3% in a year.<sup>1</sup> Head and neck schwannomas commonly present with a painless, unilateral neck mass. Occurrences of neurological symptoms like paraesthesia and neuralgic pain are rare. However, excising schwannomas without knowing their possible nerve of origin can lead to various morbidities, especially when the tumour occurs at uncommon locations, and one such morbidity is a permanent nerve injury.<sup>2</sup> Based on a study that analysed the nerve of origin of schwannomas, it can be categorised into medial and lateral groups. Lower cranial nerves of IX, X, XI, XII, and the cervical sympathetic chain consist of the medial group. Meanwhile, the lateral group comprised the tumours seen originating from the cervical plexus, cervical neck trunk, and brachial plexus.<sup>3</sup> Hereby, we present a unique case of posterior neck schwannoma in an elderly

## ABSTRACT

Schwannomas are the most commonly occurring neurogenic tumours. However, cervical plexus schwannomas are rare, and they mostly present with an asymptomatic neck mass. An elderly male with multiple comorbidities presented with a painless posterior triangle neck mass for a year without any neurological symptoms. After undergoing a few diagnostic modalities, including ultrasound-guided biopsy and MRI, the mass was suggestive of a schwannoma. Post-excision biopsy, no neurological deficits or recurrence were seen. This case exemplifies the challenges associated with diagnosing cervical plexus schwannomas and highlights the necessity of integrating multiple diagnostic modalities for their accurate identification. The rarity of this tumour, coupled with its uncommon anatomical location, highlights the importance of considering cervical plexus schwannomas in the differential diagnosis of asymptomatic lateral neck masses.

patient. We emphasise the challenges experienced in the work-up of a neck mass.

## Case report

### Clinical Presentation

A 68-year-old male with multiple comorbidities presented with a painless left-sided neck swelling that had been progressively enlarging over the past year. He did not report any obstructive symptoms, upper aerodigestive tract complaints, or constitutional symptoms such as fever, weight loss, or night sweats. There were no symptoms suggestive of pulmonary tuberculosis, and no known tuberculosis contacts.

On examination, a solitary left-sided neck mass was noted in the posterior triangle at level V, measuring approximately 3×3 cm. The mass was firm, non-tender, and not attached to the overlying skin. It was mobile in the horizontal plane but fixed in the vertical axis (Figure 1). Examination of other systems revealed no additional abnormalities.



Figure 1 Picture of the left posterior triangle neck mass.

## Investigations

### Biopsy

An initial fine-needle aspiration (FNA) yielded unsatisfactory results. Subsequently, an ultrasound-guided core needle biopsy was performed, which was reported as consistent with a schwannoma.

### Imaging

Contrast-enhanced computed tomography (CECT) of the neck demonstrated a well-defined hypodense lesion in the left posterior neck region, measuring  $2.4 \times 2.3 \times 2.7$  cm. Minimal central hypodensity was noted within the lesion, suggestive of cystic or necrotic changes. Anterolaterally, the mass was seen compressing the sternocleidomastoid (SCM) muscle and abutting the left internal jugular vein. Posteriorly and medially, it was indenting the scalene muscles and the longus capitis muscle, respectively.

Subsequent magnetic resonance imaging (MRI) revealed a T2-weighted hyperintense lesion located deep to the SCM muscle, which enhanced on post-contrast sequences. A beak-like configuration was observed pointing towards the left C2/C3 neural foramen; however, there was no widening of the foramen or intraspinal extension (Figure 2a and 2b). No brachial plexus involvement was identified. Collectively, the imaging features suggested a schwannoma arising from the cervical nerve root.

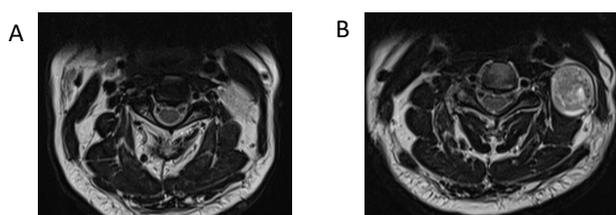


Figure 2 MRI neck axial view showing a hyperintense mass on T2-weighted, which was located deep to the SCM muscle (A), and a beak-like configuration was seen pointing towards the left C2/C3 neural foramen (B).

## Surgical Findings

The patient underwent surgical excision under general anaesthesia. Intraoperatively, a well-encapsulated mass measuring  $4 \times 2.5$  cm was visualised deep to the SCM muscle and anterior to the trapezius muscle. The greater auricular nerve and spinal accessory nerve were identified and preserved. The tumour was found to arise from the cervical plexus (Figure 3a and 3b).

Intracapsular dissection was carefully performed, and the tumour was successfully separated from the surrounding neural structures. The nerve of origin was preserved and left intact. There were no complications during the procedure.

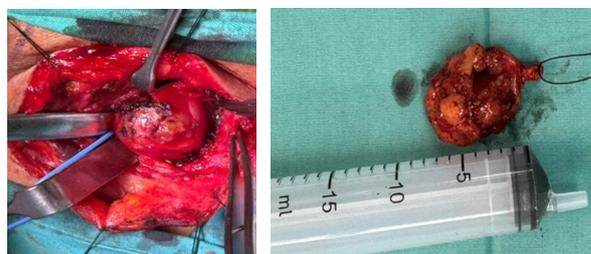


Figure 3 Intraoperative picture of the posterior neck mass (A). Gross specimen of the mass, which was excised as a whole (B).

## Histopathological Findings

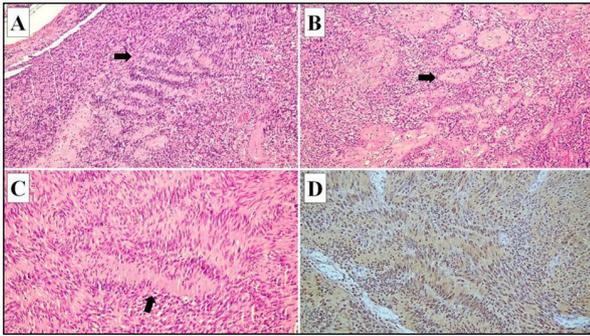
Histological examination confirmed the diagnosis of schwannoma, demonstrating classic Antoni A and Antoni B architectural patterns. Immunohistochemical staining showed diffuse positivity for S-100 protein, further confirming the diagnosis (Figure 4a–d).

## Outcome

The postoperative course was uneventful, and the patient did not develop any neurological deficits. He remains well and under regular follow-up, with no evidence of recurrence observed over the past year.

## Patient Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. Efforts have been made to ensure anonymity and confidentiality.



**Figure 4** The tumour is composed of a biphasic pattern of compact hypercellular Antoni A area (arrow) and myxoid hypocellular Antoni B area (A, H&E, x20). Focally, there are many blood vessels with thickened hyalinised walls (arrow) (B, H&E, x20). The neoplastic cells demonstrate elongated and wavy nuclei with tapered ends, and form nuclear palisading called Verocay bodies (arrow) (C, H&E, x40). These cells are positive for S100 immunohistochemistry (D, x40).

## DISCUSSION

Schwannomas are benign tumours that arise from Schwann cells, and they account for about 25%–45% of all tumours in the head and neck region.<sup>1</sup> The commonest of them are intracranial schwannomas.<sup>1</sup> However, cervical schwannomas and schwannomas originating from the cervical plexus are less commonly reported.<sup>2</sup> The cervical plexus, formed by the anterior rami of the upper four cervical nerves (C1–C4), is deeply situated beneath the sternocleidomastoid muscle. Among these, the C2 nerve root is the most frequent origin of schwannomas in this region.<sup>1</sup>

Many patients with schwannoma lack noticeable symptoms during their presentation.<sup>3</sup> Commonly, patients present with a unilateral neck mass, which is painless and slow-growing.<sup>1</sup> The challenge and difficulty in diagnosing these cases are experienced during this stage of the disease, as not many present with evidence of neurological deficit, although schwannomas are known as neurogenic tumors.<sup>3</sup> Occasionally, patients can have symptoms suggestive of Horner's syndrome, which points more towards origin from the cervical sympathetic chain.<sup>2</sup> Due to a lack of specific signs and symptoms, these benign tumours are frequently misidentified as cervical lymphadenopathies, branchial cysts, or other benign soft tissue tumours.<sup>5</sup> Zhang et al. and de Araujo et al. reported the annual tumour growing rates are 2.75mm and 3mm, respectively, supporting their indolent nature.<sup>3</sup>

Despite being a benign lesion, the risk of malignant transformation has been observed in about 8% and 13.9%

of head and neck schwannomas.<sup>3</sup> Owing to their deep anatomical location topped with subtle symptoms and minimal to absent neurological deficits, clinical suspicion alone is insufficient for its diagnosis, necessitating other investigations like imaging and histopathological evaluation.

The primary investigation for unilateral neck mass is fine-needle aspiration cytology (FNAC). It is widely available and cost-effective for evaluating lymphadenopathy and distinguishing between benign and malignant lesions.<sup>6</sup> However, FNAC has a relatively low sensitivity (0%–40%) for diagnosing schwannomas. This is due to the tumour's heterogeneous nature, which frequently undergoes cystic degeneration. Thus, they contain a combination of densely cellular areas and sparsely cellular, myxoid regions, which are Antoni A and Antoni B, respectively.<sup>7</sup> This architectural complexity contributes to the high rate of unsatisfactory FNAC results in schwannomas. FNAC also lacks both the ability to report any nerve involvement nor to use immunohistochemical analysis, such as S-100 protein to aid in its diagnostic accuracy.<sup>5,7</sup> Thus, this further reduces the role of FNAC to diagnose tumours like cervical schwannomas.<sup>8</sup> A study that analysed 30 cases of extracranial head and neck schwannomas reported only four that were able to be correctly diagnosed via FNAC. Specificity of only about 20% was detected via FNAC, which was less than imaging modalities such as CT and MRI, which had a specificity of 38%.<sup>8</sup>

Another investigation of choice is the ultrasound-guided core needle biopsy, which has emerged as a superior alternative to FNAC. A retrospective study of 154 cases of peripheral nerve sheath tumours reported an overall diagnostic accuracy of 99.3% using ultrasound-guided core needle biopsy, with sensitivity and specificity rates of 90.9% and 100%, respectively.<sup>9</sup> Core needle biopsy allows for better preservation of tissue architecture, facilitating immunohistochemical staining for markers such as S-100, SOX10, and EMA, which are key in differentiating schwannomas from other spindle cell tumours.<sup>7</sup> In our case, both FNAC and ultrasound-guided core needle biopsy were performed, with the definitive diagnosis established through core needle biopsy.

Imaging, particularly MRI, is the preferred modality for diagnosing schwannomas due to its soft tissue contrast. MRIs are particularly useful in defining tumour boundaries, evaluating involvement with adjacent nerves and vessels, and characterizing internal structure variations that are significant in head and neck tumours.<sup>1</sup> Schwannomas typically appear as well-circumscribed, encapsulated masses and are hyperintense on T2-weighted images, reflecting their biphasic cellular architecture.<sup>1</sup> Besides, diffusion-weighted MRI can aid in distinguishing benign schwannomas from their malignant counterparts as the former shows a higher apparent diffusion coefficient value (ADC) due to their lower cellular density and abundant myxoid content.<sup>10</sup> Despite MRI's diagnostic advantages, imaging alone often cannot confirm schwannomas as these tumours can mimic other soft tissue masses, thus histopathological analysis remains essential.<sup>11</sup>

Management of cervical schwannomas is primarily surgical, with extracapsular resection being the preferred approach to preserve neural function, while ensuring complete tumour excision.<sup>5</sup> However, due to the intimate association with cervical nerves, conventional extracapsular excision, while effective, carries risks of nerve injury, leading to unwanted sensory or motor deficits.<sup>11</sup> To mitigate this, intracapsular excision with gentle dissection between the tumour capsule and normal fascicles has been advocated. The epineurium layer covering the capsule should be dissected akin to peeling an onion, allowing for safe removal of the tumour via its proximal and distal poles.<sup>12</sup> Notably, fascicles within the tumour are usually non-functional, and their excision typically does not result in postoperative deficits.<sup>12</sup>

The usage of intraoperative nerve monitoring is preferred, especially for tumours originating from functionally significant nerves. Besides, the usage of an operating microscope, microsurgical instruments, and techniques is pivotal.<sup>2</sup> Advances in nerve-sparing enucleation and intraoperative neuromonitoring have significantly reduced postoperative neurological complications.<sup>5,13</sup> In our case, meticulous surgical planning and intraoperative nerve preservation strategies were employed to avoid any neurological complications.

The choice of surgical approach should be guided by the tumour's size, location, proximity to great vessels, and any suspicion of malignancy. In large or infiltrative types of schwannomas, where nerve preservation is not feasible, a nerve sacrifice may be warranted, followed by its reconstruction and rehabilitation planning.<sup>11</sup> Thorough preoperative counselling is crucial to inform patients of potential neurological sequelae. Despite their benign nature, schwannomas often require timely surgical intervention due to the possibility of progressive compression symptoms. Nonetheless, long-term surveillance is generally not necessary post-operatively given their low recurrence rates and excellent prognosis following a complete excision.

## CONCLUSION

This case exemplifies the challenges associated with diagnosing cervical plexus schwannomas and highlights the necessity of integrating multiple diagnostic modalities for an accurate diagnosis. Although neurogenic tumours like cervical plexus schwannomas are benign in nature, they should be considered as part of the differential diagnosis of any unilateral, slow-growing neck mass in adults.

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