

Middle-aged woman with painless neck swelling

A 57-year-old woman presented with a 2-month history of an asymptomatic left neck lump. She is a non-smoker, does not drink alcohol, and has no family history of head and neck cancers. On examination, there was a 2cm left cervical level II ovoid and mobile nodule, which appeared to exhibit transmitted pulsations. Cranial nerve examination was normal nasoendoscopic and otoscopic evaluation were unremarkable.

The lesion was visualised on both computed tomography and magnetic resonance imaging (MRI) scans as a well-defined 2.8 x 2.9 x 2.8cm mass just adjacent to the major vessels in the neck (Fig. 1).

What is the diagnosis?

- A. Enlarged cervical lymph node
- B. Carotid body tumour
- C. Vagal schwannoma
- D. Sympathetic chain schwannoma
- E. Branchial cleft cyst

The patient then underwent surgical excision of the lesion. Histological examination revealed a dense lesion with adjacent compressed nerve fibres containing ganglions that displayed a mixture of both cellular Antoni A and hypocellular Antoni B segments, with the latter showing vessels with surrounding hyalinisation. These findings are consistent with a schwannoma—in this case, a sympathetic chain schwannoma.

Schwannomas are nerve sheath tumours consisting of Schwann cells that confer a low risk of malignant

transformation,¹ of which 25-45% are found in the head and neck region.² Schwannomas of cervical sympathetic chain (SCSC) are even more uncommon³ with fewer than 80 cases reported in the literature, of which only 12 cases had preoperative Horner's syndrome.⁴ The sympathetic chain is a longitudinal structure lying bilaterally adjacent to the spine, extending from the base of the skull to the coccyx with cervical, thoracic and lumbar components. It is part of the autonomic nervous system that takes part in the “fight or flight” response, which includes pupillary dilatation, perspiration and tachycardia. The cervical segment provides innervation to orbital structures (pupil dilatory muscle and Müller's muscle) and facial sweat glands, as well as contributes branches to the cardiac plexus.

Pre-excision symptom complexes are usually related to compressive effects of large tumours, for example, dysphagia, dyspnoea, and cranial nerve (CN) deficits (CN X, XII).⁴ However, patients often present with an asymptomatic neck swelling for which the main differentials are vagal schwannomas (most common), carotid body tumours and schwannomas from surrounding cranial nerves (glossopharyngeal and hypoglossal).

Schwannomas typically appear heterogeneously bright on T2-weighted images due to the presence of interspersed Antoni A (hypercellular) and Antoni B (hypocellular) tissue signals.⁵ Occasionally, a split-fat sign can also be appreciated (Fig. 1B arrowhead)—it describes a thin rim of peripheral fat seen



Fig. 1. (A) T1-weighted fat-suppressed axial magnetic resonance imaging (MRI) with contrast. (B) T1-weighted coronal MRI non-fat sat image. (C) Magnified T1-weighted MRI axial cuts showing relation of the lesion with major vessels.

* External carotid artery; # internal carotid artery; ^ internal jugular vein

Answer: D

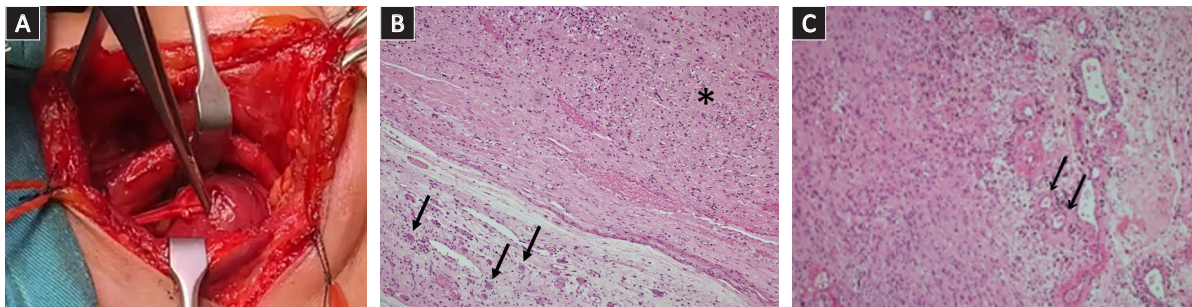


Fig. 2. (A) Intraoperative schwannoma with attached normal sympathetic chain nerve fibres inferiorly. (B) Schwannoma (*) showing a well-demarcated border from the adjacent compressed nerve tissue which contains ganglion cells (arrows) (haematoxylin and eosin [H&E] staining, 100x magnification). (C) Cellular Antoni A area (left) and hypocellular myxoid Antoni B area (right). The Antoni B area also contains vessels with surrounding hyalinisation (arrows) (H&E staining, 100x magnification).

especially on coronal cuts, suggesting a nerve sheath tumour.⁶

Due to the posteromedial location of the cervical sympathetic chain in relation to carotid sheath, it is characteristic of SCSC to:⁷

- (1) displace the internal carotid artery (ICA) anterolaterally, unlike vagal schwannomas that medialise the ICA;
- (2) not splay the ICA and internal jugular vein (IJV), as opposed to vagal schwannomas that characteristically

do, due to their position within the carotid sheath between the carotid and IJV.

However, it must be acknowledged that the above features are not uniformly seen in all cases, and variations have been observed in literature.⁸ In a study by Anil and Tan,⁸ one SCSC demonstrated posterior displacement of the ICA while another had the SCSC splaying the ICA (anteriorly) and IJV (posteriorly), although this is rarely seen. It is important to differentiate between vagal and sympathetic chain schwannomas, as excising the former

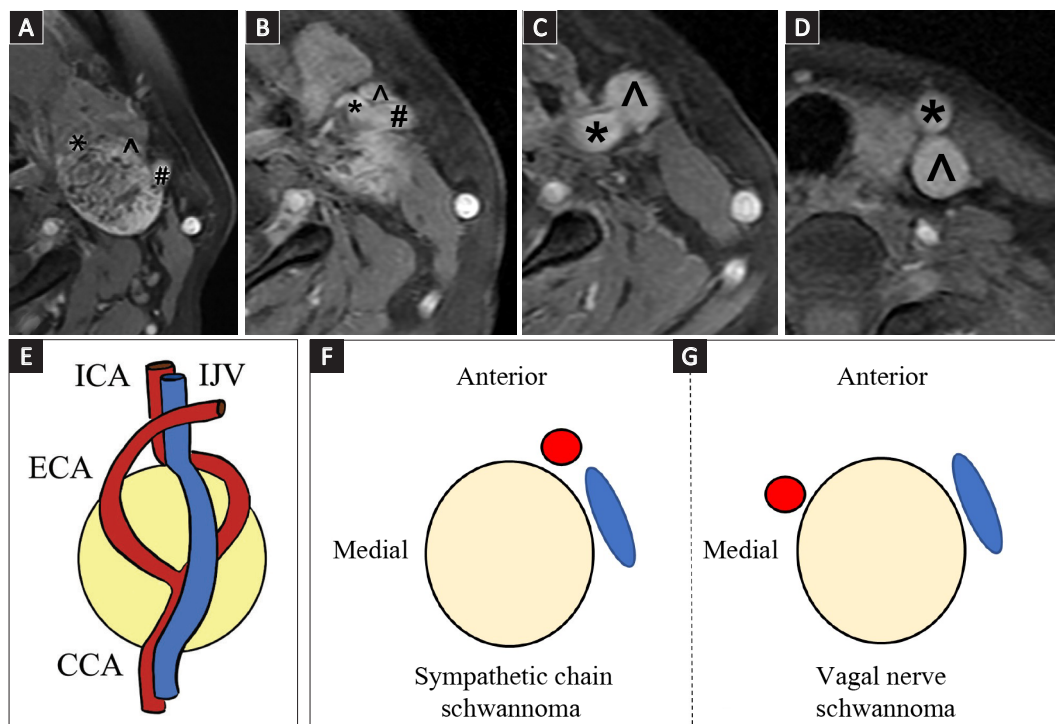


Fig. 3. Course of internal jugular vein (IJV) and carotids from superior to inferior. (A) Displacement of IJV and internal carotid artery (ICA) anterolaterally with splaying of carotids. (B and C) IJV compressed anteriorly at level of carotid bifurcation and immediately inferiorly. (D) Common carotid artery (CCA) crosses IJV and appears anterior to the vein at the level of the thyroid gland. (E) Diagram depicting altered anatomy found in this case study where the schwannoma caused displacement and intertwining of the ICA, external carotid artery (ECA) and IJV. (F and G) Schematic diagrams showing the expected relations between the structures in the cases of a sympathetic chain schwannoma, (F) or vagal nerve schwannoma.

* ECA/CCA; # ICA; ^ IJV

Large circle: lesion; small circle: ICA; flat oval: IJV

will result in more significant functional deficits such as dysphonia, dysphagia, and even dyspnoea or asphyxiation from bilateral vocal cord palsy if the patient has existing contralateral vocal cord pathology.

Paragangliomas, such as carotid body tumours, classically show a “salt-and-pepper” appearance. “Salt” represents haemorrhage presented as hyperintense foci seen on T1-weighted images, while “pepper” are vascular flow voids which are hypointense on T1/2 but should have corresponding linear vascular enhancement on post-contrast images to be true vascular flow voids. Both of these are not demonstrated in this patient’s scans. Although the lesion does splay the carotid arteries superiorly, it does not demonstrate this feature exactly at the level of the carotid bifurcation (lyre sign), which is the most common location for carotid body tumours. Branchial cleft cysts will also appear markedly hyperintense on T2-weighted images due to the high fluid content.

SCSC can be managed both conservatively or surgically. The usual indications for excision of a schwannoma are: a large or fast enough growing lesion that causes problems as a result of compression of surrounding structures; or a relatively fast-growing lesion on interval scan in a younger patient, such that it is anticipated that the lesion will cause functional problems (e.g. Horner’s syndrome) during the lifespan of the patient. Conservative management can be considered in an asymptomatic patient with a stable lesion, who is able to confidently identify red flag symptoms, and comply with regular surveillance scans. In this case, the patient was recommended conservative management with serial scans. However, she was very keen for excision due to derived stress from her condition and concerns regarding future potential complications and inability to tolerate surgeries with advanced age.

Patients also need to be counselled on the development of post-excision Horner’s syndrome: ptosis, miosis and anhidrosis. They should also be informed of the risks of bleeding from vessel injury and adjacent cranial nerve palsies, potentially impacting swallowing, speech and articulation; and shoulder weakness.

Also, surgical exposure would likely involve significant manipulation of the ICA and thus, the carotid sinus. Carotid sinus manipulation intraoperatively has been reported to cause hypersensitivity, which may manifest as sinus bradycardia and third-degree heart blocks, more so in the elderly population.⁹ As such, injection with 2mL of 1% lignocaine into the adventitia at the carotid bifurcation should be performed prior

to manipulation for hydrodissection and to reduce the sensitivity of the carotid sinus body.¹⁰

In our patient, postoperatively, she had preserved palatal elevation, swallowing function, shoulder shrug and no tongue deviation on protrusion. Signs of Horner’s syndrome were evident with left anhidrosis and miosis, but symmetrical palpebral aperture was still preserved. Two months postoperatively, the patient went on to develop left partial ptosis due to weakness of Müller’s muscle and superior tarsal muscle, which completed the triad. As of her last clinic review, her ptosis has improved slightly.

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