

CASE STUDY

# A Report on a Case and Literature Review on Cervical Sympathetic Chain Schwannoma that Mimics a Paraganglioma

Youssef Lakhdar<sup>1</sup>, Youssef El Khalifa<sup>1,\*</sup>, Aitlhadj Mohamed Amine<sup>1</sup>,  
Othman Benhoummad<sup>2</sup>, Youssef Rochdi<sup>1</sup>, and Abdelaziz Raji<sup>1</sup>

## ABSTRACT

**Background:** Cervical sympatric schwannoma is a rare benign tumor originating from de Schwan cells. Its clinical presentation is misleading and it can remain asymptomatic for a long time. Mostly, it appears as slow slow-growing cervical mass and may be a differential diagnosis with other neck masses such as paragangliomas. We discuss a case of Cervical sympatric schwannoma and go over its radiological, surgical, and surgical characteristics.


**Case Presentation:** With no prior medical history, a 61-year-old lady appeared with a right cervical mass that had been present for over two years but had lately grown in size without causing dysphagia, dysphonia, or dyspnea. Clinical examination found a firm, slightly pulsatile, non-painful latero-cervical mass measuring 2 × 3 cm. Without other abnormalities on examination of the upper aerodigestive tract or thyroid gland and no neurological deficit. Cervical Ultrasound showed an oval tissular mass with regular contours in close contact with the right common carotid artery. Computed Tomography showed a right superior jugulocarotid mass located behind the vascular axis of the neck with early central vascularization. On magnetic resonance imaging (MRI), this mass was well limited, hypointense on T1 and hyper-intense on T2 with intense contrast enhancement, corresponding to a paraganglioma or schwannoma. A full blood work came back negative. Our patient underwent a complete surgical excision of the tumour which originated in the deep branches of the cervical plexus. Following surgery, the patient had Horner's syndrome, histopathology confirmed the diagnosis of schwannoma.

**Conclusions:** It is still challenging to differentiate between cervical sympathetic chain schwannoma and paraganglioma clinically and radiologically. In the case of the schwannoma surgical approaches must be as conservative as possible to avoid damages of the origin nerve and the patient must be aware of the possibility of Horner syndrome.

**Keywords:** Paraganglioma, Schwannoma, Sympathetic Nervous System.

Submitted: February 08, 2024

Published: April 08, 2024

 10.24018/ejclinimed.2024.5.2.331

<sup>1</sup>ENT and HNS Surgery Department, Mohammed VI University Hospital, Morocco.

<sup>2</sup>Faculty of Medicine and Pharmacy, Ibn Zohr University, Morocco.

\*Corresponding Author:

e-mail: elkhalifayoussef@gmail.com

## 1. INTRODUCTION

Upper cervical schwannoma, previously called neurilemmoma, is a rare benign tumor, originating from the Schwan cell. It only made up 10% of schwannomas cases, and most of the people who develop it are between the ages of 40 and 50 [1]. This benign tumor involves the pneumogastric nerve mainly [2], schwannoma deriving from the cervical sympatric chain is very rare [3] with

less than 50 cases reported in the literature [4]. Clinical presentation is misleading, it is usually an asymptomatic slow-growing cervical mass. The diagnosis is based on radiological findings, especially CT scan and IMR [5]. However, differential diagnosis with other neck masses such as paraganglioma may be difficult to establish and intraoperative exploration may be required.



## 2. PATIENT AT OBSERVATION

A 61-year-old lady who had no prior medical or surgical history appeared with a right cervical tumor that had been growing for the previous two years, without signs of cervical compression (no dysphagia, dysphonia or dyspnea) and progressing in the context of apyrexia and preserved general condition. A 2 × 3 cm isolated superior right lateral cervical tumor was discovered during a physical examination, which was oblong, firm, slightly pulsatile, and insensitive. This mass was mobile horizontally but not vertically. The skin around was normal. There was no associated cervical adenopathy or other abnormalities on ENT examination. The neurological examination revealed no abnormalities.

Cervical Ultrasound showed a right superior jugulo-carotid tissular mass in close contact with the right common carotid artery. This tumour was measuring 31 × 18 mm, was oval, well limited, hypoechogenic and heterogeneous. On the CT scan the mass was isodense, well-limited, located medially to the sternocleidomastoid muscle and coming in contact with the posterior surface of the carotid artery and the internal jugular vein which remains permeable with early central contrast enhancement suggesting a diagnosis of paraganglioma or shwannoma (Fig. 1). On MRI the tumor was hypointense in T1-weighted images with high contrast enhancement, and discreetly heterogeneous hyperintense in T2-weighted images, respecting neighboring structures (Fig. 2). Biological and radiological assessment for multiple endocrine neoplasia came back negative. The tumour was approached surgically under general anesthesia to determine its nature and origin via a modified S  bileau-Carrega incision. After freeing the sternocleidomastoid muscle and vessel-loop control of the right common carotid, and the internal jugular vein, the initial part of carotid arteries, both internal and external and after vagus nerve dissection, surgical exploration revealed a homogeneous oval yellowish retrovascular tumor measuring 4 × 3 cm, moving the carotid artery and internal jugular vein anteriorly and the pneumogastric nerve without invading them (Fig. 3). It seemed to be coming from the cervical plexus's deep branches, the excision of the tumor was total with a sacrifice of the nervous branch of origin with no bleeding. Postoperatively the patient presented a Horner's syndrome (reactive right unilateral media) with no other complication, thus confirming the origin to be a sympathetic chain. Histopathological analysis of the tumour was consistent with a benign schwannoma. No recurrence after 6 months of follow-up.

## 3. DISCUSSION

*Neurilemmomas* was the term used in 1935 to describe tumours originating from nerve sheath elements [6]. Numerous terminologies for schwannomas are used in the literature, which has historically complicated histological interpretation and postponed an accurate diagnosis [7]. The head and neck areas are where twenty to forty-five per cent of all extracranial schwannomas occur [8]. The clinical presentation lacks specificity, in most cases, it's a

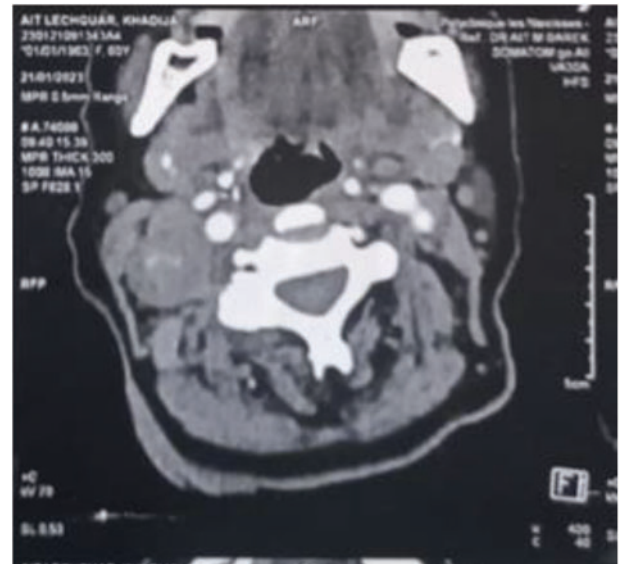


Fig. 1. Cervical CT scan in the axial section after injection of contrast medium, showing a right jugular-carotid mass measuring 23 × 27 mm with early central vascularisation in favour of a glomus tumour or schwannoma.

neck mass slowly gaining in volume over a long period. Pharyngeal discomfort and odynophagia may be possible due to pharyngeal compression. An association of neck pain and signs of neurological deficit are suggestive of malignancy [9].

Fine needle aspiration may be a helpful tool to confirm the diagnosis, but it's considered risky when a paraganglioma is suspected.

CT scan and MR imaging are useful for determining the characteristics of the tumour, in particular its location, size and contours, but also risks, in order to plan the surgical procedure. Schwannomas often manifest as well-delimited tumors that push aside surrounding structures without infiltrating them [10]. On MRI, they usually exhibit low T1 signal, high T2 signal, and significant contrast enhancement. On angiography, they are correspondingly avascular or hypovascular. Radiologically, schwannoma poses a differential diagnosis problem with cervical paraganglioma, given the intense enhancement after contrast injection of shwannoma because of the accumulation of contrast medium in the interstitial spaces [11]. Other paraganglioma manifestations that may be useful in differentiating the two entities include the classic "salt & pepper" flow gaps on MRI, as well as a heterogeneous enhancement pattern brought on by isolated thrombi or hemorrhage in large lesions [12]. Schwannomas may show signs of vascularity within the tumour or around the capsule [13], their density is smaller than that of the nearby muscles; Since intra-tumoral capillaries are primarily responsible for vascularization following injection, late enhancement is often uniform. Reference [14], As has been previously shown, certain cervical schwannoma can show significant enhancement on computed tomography and magnetic resonance imaging (MRI) even though the condition is often hypovascular. The hypovascular character of these lesions and the corresponding inadequate venous drainage of contrast appear to be the cause [15].

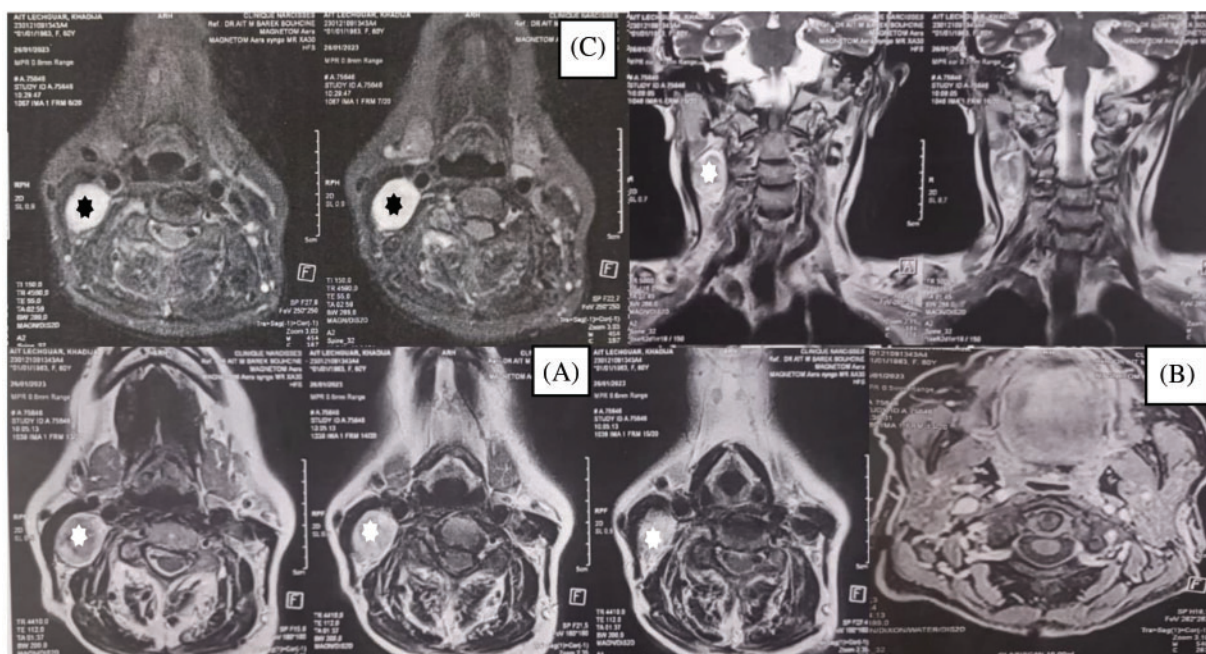


Fig. 2. MRI cervical showing a right laterocervical mass in close contact with the carotid primitive, hyposignal T1 (A), discretely heterogeneous hypersignal T2 (B), intensely contrast-enhanced (C)\*.

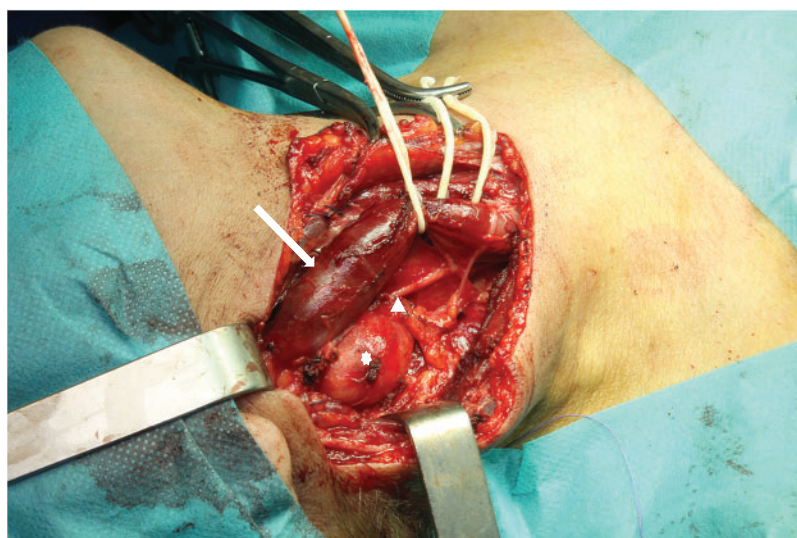


Fig. 3. Perioperative view showing a homogeneous oval yellowish tumor (asterisk), retrovascular and originating from the cervical plexus's deep branches (triangle), the internal jugular vein displaced anteriorly (arrow), the carotid and the pneumogastric nerve.

Near the carotid bifurcation, schwannomas originating from the cervical sympathetic chain and lower four cranial nerves can also occur. Before surgery, a schwannoma originating from the cervical sympathetic chain may resemble a preoperative carotid body tumor (CBT) and cause the bifurcation to splay [15]. Therefore, accurate preoperative Schwannoma differential diagnosis and CBT are critical for surgical planning and postoperative problems prediction. The course of treatment would change if CBT was suspected, and preoperative embolization would probably be required [16]. Other differential diagnoses include inflammatory or metastatic adenopathies.

The therapeutic attitude via neck schwannomas is surgery primarily [17]. Usually, the nerve is sacrificed during the excision. Then, the most common postoperative consequence is Horner's syndrome. It is imperative to preserve the nerve of origin owing to the benign nature

of lesions. Additionally, a conservative approach needs to be taken into consideration given the non-invasive nature, moderate development, and extremely low recurrence rate [17]. To properly detect and categorize nerve sheath tumors, electron-microscopy and immunohistochemical studies (S-100, Leu-7) are required [18].

#### 4. CONCLUSION

Schwannoma should be included in the differential diagnosis and CBT options when dealing with the cervical sympathetic chain, sometimes radiological findings may not be able to differentiate between the two. The main difference is the lack of intensity, the surgical approaches must in all cases be as conservative as possible to avoid

damage to nerve damage and the patient must be aware of the possibility of Horner syndrome.

#### CONFLICT OF INTEREST

Authors declare that they do not have any conflict of interest.

#### REFERENCES

- [1] Airlangga PA, Prijambodo B, Hidayat AR, Benedicta S. Schwannoma of the upper cervical spine—a case report. *Chin J Traumatol*. 2019 Dec;22(6):368–72. doi: 10.1016/j.cjtee.2019.07.005.
- [2] Bozec A, Dassonville O, Poissonnet G, Niadje M, Demard F. Laryngeal schwannoma: a case report. *Ann Otolaryngol Chir Cervicofac*. 2003;120(1):40–4.
- [3] Colreavy MP, Lacy PD, Hughes J, Bouchier-Hayes D, Brennan P, O'Dwyer AJ, et al. Head and neck schwannomas—A 10 year review. *J Laryngol Otol*. 2000;114(2):119–24.
- [4] Yim DW, Sheridan MF. Cervical sympathetic schwannoma: a case report and review of the English literature. *Otolaryngol Head Neck Surg*. 1997;117:S206–10.
- [5] Manzanedo-Romero I, García-Muñoz-Najar A, Acín-Gándara D, Carrión-Álvarez L, Urbasos-Pascual M, Pereira-Pérez F. Cervical sympathetic chain schwannoma associated with parathyroid adenoma. *Cirugía Y Cirujanos*. 2015 September–October;83(5):409–13.
- [6] Stout A. Peripheral manifestations of the specific nerve sheath tumour (neurilemmoma). *Am J Cancer*. 1935;24(4):751–80.
- [7] Baillet JW, Abemayor E, Andrews JC, Rowland JP, Fu YS, Dawson DE, et al. Malignant nerve sheath tumours of the head and neck: a combined experience from two university hospitals. *Laryngoscope*. 1991 Oct;101(10):1044–9.
- [8] Castelijns JA, Snow GB, Leverstein H. The value of magnetic resonance imaging in the differential diagnosis of parapharyngeal space tumours. *Clin Otolaryngol*. 1995 Oct;20(5):428–33.
- [9] Elias MM, Balm AJ, Peterse JL, Keus RB, Hilgers FJ. Malignant schwannoma of the parapharyngeal space in Von Recklinghausen's disease; a case report and review of the literature. *Laryngol Otol*. 1993 Sep;107(9):848–52.
- [10] Casserly P, Kiely P, Fenton JE. Cervical sympathetic chain schwannoma masquerading as a carotid body tumour with a postoperative complication of Worst-bite syndrome. *Eur Arch Otorhinolaryngol*. 2009 Oct;266(10):1659–62.
- [11] Van den Berg R, Schepers A, de Bruïne FT, Liauw L, Mertens BJA, Van der Mey AGL, et al. The value of MR angiography techniques in the detection of head and neck paragangliomas. *Eur J Radiol*. 2004 Dec;52(3):240–5.
- [12] Som PM, Braun IF, Shapiro MD, Reede DL, Curtin HD. Tumors of the parapharyngeal space and upper neck: MR imaging characteristics. *Radiol*. 1987 Sep;164(3):823–9.
- [13] Aygenc E, Selcuk A, Ozdem C. Hypervascular parapharyngeal schwannoma: an unusual case. *Auris Nasus Larynx*. 2002 Apr;29(2):215–7. doi: 10.1016/s0385-8146(01)00130-4.
- [14] Nao E-E-M, Dassonville O, Bozec A, Sudaka A, Marcy P-Y, Vincent N, et al. Cervical sympathetic chain schwannoma. *Eur Ann Otolaryngol, Head Neck Surg*. 2012 Feb;129(1):51–3.
- [15] Tomita T, Ozawa H, Sakamoto K, Ogawa K, Kameyama K, Fujii M. Diagnosis and management of cervical sympathetic chain schwannoma: a review of 9 cases. *Acta Oto-Laryngol*. 2009 Mar;129(3):324–9. doi: 10.1080/00016480802179735.
- [16] Wax MK, Shiley SG, Robinson JL, Weissman JL. Cervical sympathetic chain schwannoma. *Laryngoscope*. 2004 Dec;114(12):2210–3. doi: 10.1097/01.mlg.0000149460.98629.42.
- [17] Myssiorek DJ, Silver CE, Valdes ME. Schwannoma of the cervical sympathetic chain. *J Laryngol Otol*. 1988 Oct. doi: 10.1017/S0022215100106930.
- [18] Donnelly MJ, Al-Sader MH, Blayney AW. View from beneath: pathology in focus. Benign nasal schwannoma. *J Laryngol Otol*. 1992 Nov;106(11):1011–5. doi: 10.1017/s0022215100121644.