



Case Report

Schwannoma of the sympathetic trunk: A case report

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ABSTRACT

Introduction and Importance: Schwannomas are benign tumors commonly found in the cranial vault at the cerebellopontine angle. Schwannomas could arise from any nerve that has Schwann sheath. Their most common extra-cranial localization is the parapharyngeal space. Schwannomas do not usually metastasize, and recurrence is uncommon.

Case presentation: We present a case of a sympathetic trunk schwannoma in a patient that presented with dysphagia and a painful neck mass. Oro-pharyngeal examination revealed the presence of a mass filling-up the left tonsillar fossa, and pushing the uvula to the right side. The larynx was also deviated to the right due to the mass effect. The mass was resected en-bloc with the involved part of the sympathetic trunk. The patient developed Horner's syndrome postoperatively.

Clinical discussion: The occurrence of Sympathetic trunk Schwannomas is very rare. The majority of patients presented with a cervical mass and non-specific symptoms. The most effective treatment is surgical resection. Our experience suggests sympathetic trunk Schwannomas as a differential diagnosis of slow-growing neck masses and asserts that the surgical resection is the main treatment.

Conclusion: The occurrence of Schwannomas in the sympathetic trunk is rare. High clinical suspicion is required to achieve the preoperative diagnosis. The optimal management is the total surgical resection. Recurrence is uncommon when the mass is totally resected.

1. Introduction

Schwannomas are commonly found in the cranial vault at the cerebellopontine angle. Schwannomas could also be found in extra-cranial localizations in the head and neck region [1]. Their most commonly reported extra-cranial localization is the parapharyngeal space [2]. When Schwannomas are located in the neck, they could mimic the carotid body tumors [3].

Schwannomas of the head and neck form a small percentage of the head and neck neoplasms. Schwannomas could arise from any cranial autonomic or somatic nerve that has Schwann sheath.

In the neck, Schwannomas arise either medially or laterally. Medial localizations of the cervical Schwannomas could indicate that they arise from the sympathetic trunk, or from the last four cranial nerves [4]. The majority of cervical Schwannomas neck originate from the sympathetic trunk and the Vagus nerve [5,6].

Schwannoma could occur solely, or could be related to neurofibromatosis type 2 (NF2) [7].

The clinical presentations of cervical Schwannomas are numerous [8], when compared with carotid body tumors which present as asymptomatic gradually growing mass in the neck [9]. In isolated lesions, malignant transformation is very rare [7]. In addition, it is well-known that Schwannomas do not metastasize [10]. Recurrence is uncommon when en-bloc resection is performed [11].

To the best of our knowledge, only 50 cases of schwannoma of the sympathetic trunk have been reported in the medical literature [12].

In this case, we describe the diagnostic work-up and surgical management of a sympathetic trunk schwannoma that was found in 29-years-old man.

This case report has been reported in line with the SCARE Criteria [13].

2. Case presentation

A 29-year-old man presented at the outpatient clinic of our university hospital complaining of dysphagia and a painful lump in the left part of

Abbreviations: MRI, magnetic resonance imaging; CT scan, Computed tomography scan; US, ultrasound.

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his neck. The patient also reported that he suffered from intermittent night sweats without any significant weight loss during the previous months. His medical history revealed that he suffers from deafness and dumbness since childhood. His surgical history was remarkable for varicocele and tonsillectomy that were performed 7 and 3 years ago, respectively. But family medical history, psychological history and drugs were unremarkable. Upon physical examination, his vital signs were within the normal limits. A palpable painful mass in the neck at the level IB was noted. The mass was fixed on the depth but the skin was moving on it. On lymph node examination, palpable cervical adenopathy was unremarkable. Oro-pharyngeal examination revealed the presence of a mass filling-up the left tonsillar fossa and the left part of the oropharynx, pushing the uvula to the right side. The nasopharynx, larynx, and the movement of the vocal cords were unremarkable upon the fibro-optic naso-laryngoscopy. However, the larynx was slightly deviated to the right due to the mass effect.

Magnetic resonance imaging (MRI) of the head and neck revealed a heterogeneous well-defined mass measuring 4×5 cm in the left parapharyngeal space. The mass appeared to contain numerous foci with high signal intensity on T2-weighted MRI. The compression of the oropharynx and laryngopharynx by the mass was also obvious (Fig. 1).

Contrast-enhanced computed tomography (CT) scan was performed to rule out any potential metastasis, which was completely normal. Based on these findings, we decided to remove the tumor surgically.

Under general anesthesia, an incision was made below the level of the left submandibular gland. The gland was resected and the incision was extended posteriorly towards the sternocleidomastoid muscle, and anteriorly towards the lower lip. The carotid sheath was dissected and isolated. We noticed that the locations of the carotid artery and the internal jugular vein were inverted. The mass was pushing the carotid artery posteriorly and pushing the jugular vein anteriorly. The external carotid artery was ligated above the level of the branching point of the superior thyroid artery to control any potential source of hemorrhage. The digastric and stylohyoid muscles were cut and the route of the XII cranial nerve was isolated. The para-median mandibulotomy was done, and the floor of the mouth was incised to perform the parapharyngeal dissection in order of reaching the upper borders of the mass via transcervical transmandibular swing approach. The lingual artery was also ligated. When the tumor was reached, it was found to deeply involve the sympathetic trunk (Fig. 2). Therefore, to achieve full resection of the tumor, and to prevent future recurrence, the mass was resected en-bloc with the involved part of the sympathetic trunk. The removed bone flap was returned and fixed in its place using wires. The patient's recovery was uneventful. The surgery took 5 h and no difficulties were encountered.

Gross examination of the resected specimen showed a greyish well-circumscribed encapsulated mass measuring 40 × 50 mm. Microscopic examination showed proliferation of spindle Schwann cells in biphasic compact hypercellular areas (Antoni A) and hypocellular areas (Antoni

B)(Fig. 3:A,B,C). whereas, it was diffuse and strong positive immunoreactivity for S100 (Fig. 3-D). On this basis, a sympathetic trunk Schwannoma was diagnosed.

After a 3-month period of clinical follow up, the patient's symptoms were completely resolved. Although the patient developed Horner's syndrome in the left side of his face, this had minimal effects on his life style compared to his previous complaints.

3. Discussion

Schwannomas of the head and neck are extremely uncommon, accounting for 25–45 % of the overall localizations in the body [8,14].

These neoplasms are often benign and malignant transformation is almost non-existing [7]. As a result, Schwannomas do not metastasize [10], and after en-bloc resection, recurrence is uncommon [11]. Schwannomas generally vary in size [12].

In the extra-cranial localizations of the head and neck, most authors reported that the Vagus nerve and the sympathetic trunk to be the origin of the Schwannoma.

According to a previous study, patients could present with an asymptomatic palpable mass, nerve palsy (e.g. Horner's syndrome, vocal cord palsy), pain and obstructive symptoms. In our case, the patient presented with a cervical mass and dysphagia.

Additionally, the diagnosis of Schwannoma could be done according to the clinical presentations and radiologic imaging findings [15].

A study showed that 70 % of schwannomas appear as a low intensity rim on MRI. In other words, on T1-weighted images the mass is not always clear but on T2-weighted images it tends to show increased signal intensity. After gadolinium injection, small Schwannomas look homogenous whereas larger masses tend to be heterogenous, similar to the presented case.

When Compared to MRI, CT scan reveals a well-defined mass without cystic or necrotic areas [16].

Moreover, Yasumatsu et al. reported that the sensitivity of ultrasound was very low [17]. Besides, it was reported that biopsies are not useful in achieving the diagnosing [18]. Fine needle aspiration cytology does not have satisfactory sensitivity and specificity in diagnosing the Schwannoma [19,20].

The surgical management is considered the most effective treatment to avoid recurrence [1]. Otherwise, the surgical resection of the tumor and associated neural structures may affect the patient's life style. Many patients develop Horner's syndrome after surgery [14], similar to our case. According to a study, the authors did not prefer using grafts to reconstruct the damaged part of the nerve, and suggested meticulous microsurgical techniques as the most effective solution to preserve the sympathetic trunk and prevent Horner's syndrome [12]. For large Schwannomas, it was found that radiation may be required when the mass causes serious problems [10].

In this case, the most reliable treatment was the total surgical

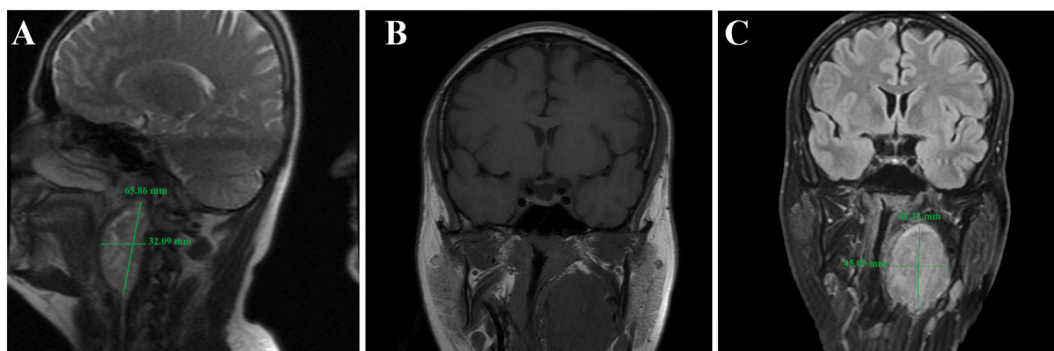


Fig. 1. Magnetic resonance imaging of the head and neck demonstrating the localization of the mass in the parapharyngeal space. A, B: T1-weighted MRI. C: T2-Weighted MRI.

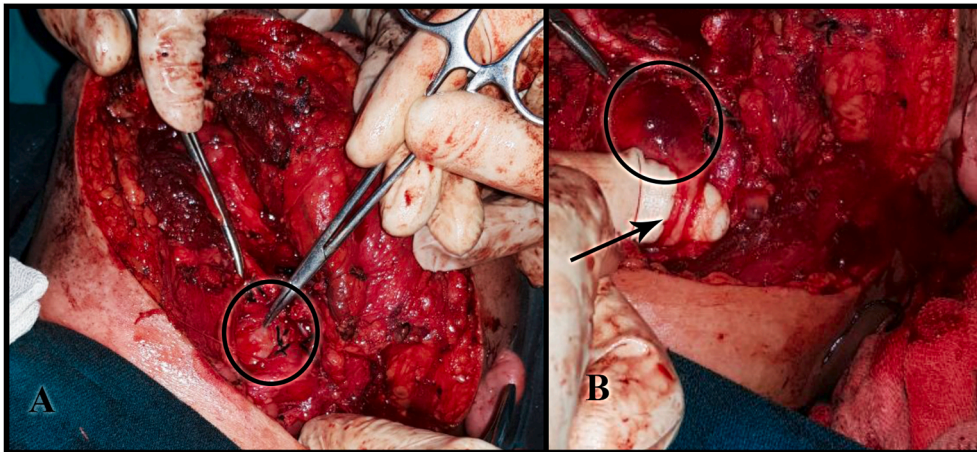


Fig. 2. The sympathetic trunk Schwannoma in situ. Note the close involvement with the sympathetic trunk.

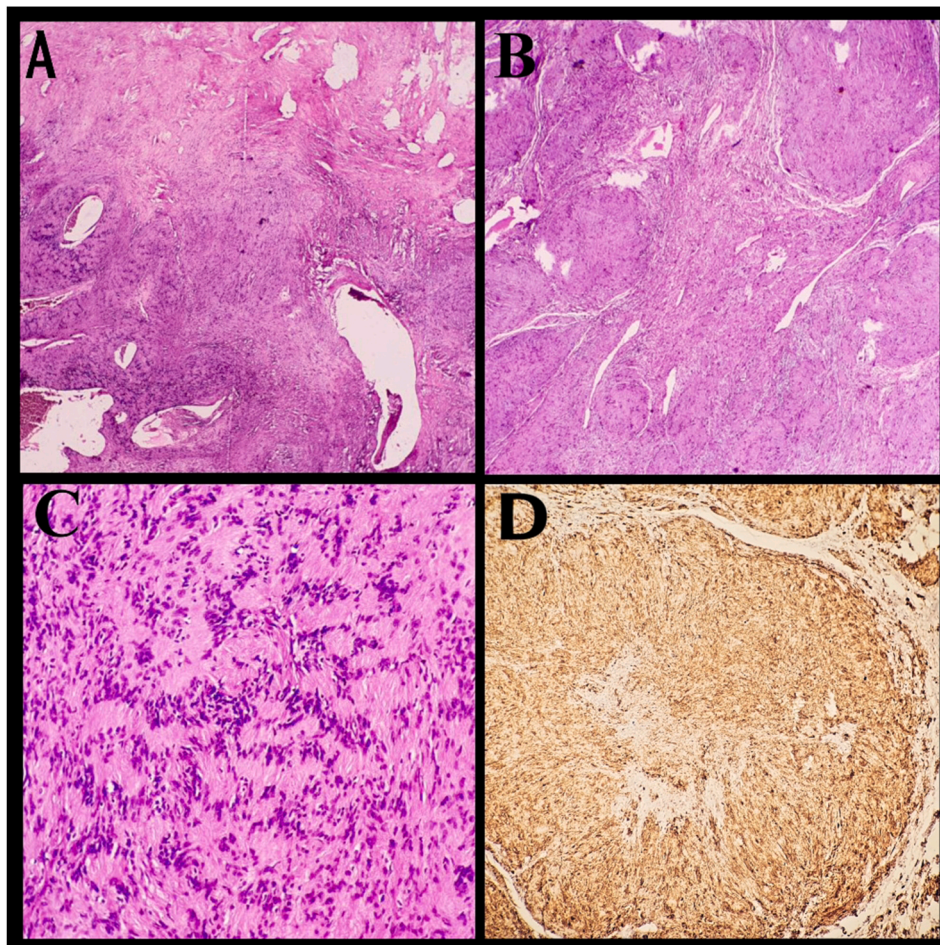


Fig. 3. A) Hyalinized stroma with variably sized ectatic blood vessels (Hematoxylin and eosin stain $\times 40$), B) Cellular area alternating with hypocellular areas (Hematoxylin and eosin stain $\times 40$), C) Cellular areas showing nuclear palisading and verocay bodies (Hematoxylin and eosin stain $\times 200$), D) Diffuse and strong positive immunoreactivity for S100 (immunohistochemically, $\times 100$).

resection, despite developing Horner’s syndrome postoperatively. However, appropriate patient counseling and balancing between the risks and benefits of the excision is necessary. Generally speaking, we suggest avoiding the excision of asymptomatic incidentally-diagnosed Schwannomas. But we learned from our case that occasionally microsurgical excisional techniques is the most optimal management for sympathetic trunk Schwannomas cases, and the absolute diagnosis could

not be reached until an exploratory surgery is done.

4. Conclusions

Schwannomas of the sympathetic trunk are rare and difficult to be diagnosed. The most common symptom is a slow-growing cervical mass. Large Schwannomas could provoke various symptoms due to the mass

effect. Radiologic imaging findings are not diagnostic but MRI and CT scan may be useful to define the mass. The best management of such cases is the total surgical resection. Recurrence is uncommon when the mass is totally resected.

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All the authors declared that they have no conflicts of interest.

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Ethical approval

Informed consent was taken for this case report. Our study ethical aspects were reviewed and approved by Damascus University deanship, Damascus, Syria.

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Author contribution

Basel Al-Ghotani: Reviewed the literature, wrote the article's abstract, introduction, and part of discussion. Bana Abo-Shdeed: Reviewed the literature, wrote the case presentation, and part of the discussion. Areej Alassaf: Led the surgery and supervised the scientific and academic aspects of the manuscript preparation and submission. All the authors read and approved the content of this manuscript.

Patient's consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Availability of data and materials

All the relevant patient data and clinical history is provided within this article.

Conflict of interest

All the authors declared that they have no conflicts of interest.

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Appendix A. Supplementary data

Supplementary data to this article can be found online at <https://doi.org/10.1016/j.amsu.2021.102624>.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Registration of research studies

1. Name of the registry: this case report is not a first time of reporting, new device or surgical technique. So I would not need a Research Registry Unique identifying number (UIN).
2. Unique Identifying number or registration ID:
3. Hyperlink to your specific registration (must be publicly accessible and will be checked): It is a case report not a research.

Guarantor

The Guarantor is the one or more people who accept full responsibility for the work and/or the conduct of the study, had access to the data, and controlled the decision to publish Mr Basel Al-Ghotani.

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