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A Rare Case of Vagus Nerve Schwannoma Presenting as a Neck Mass

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Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
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Conflict of interest: None declared

Patient: Female, 55
Final Diagnosis: Left vagal nerve schwannoma
Symptoms: Dysphagia
Medication: —
Clinical Procedure: Surgery
Specialty: Internal Medicine

Objective: Rare disease

Background: Vagus nerve schwannoma is a benign neoplasm that usually presents as an asymptomatic slow growing mass, and its presentation as a neck mass is rare. The diagnosis can be difficult to make and complete surgical excision is challenging due to the proximity of the vagus nerve fibers from which it originates. The most common symptom associated with vagus nerve schwannoma arising in the neck is hoarseness due to vocal cord palsy.

Case Report: We report a case of a 55-year-old woman who presented to the clinic complaining of throat irritation and feeling of something stuck in her throat for the past three months. On examination, a bulging left parapharyngeal mass was noted, displacing the left tonsil and uvula medially. A contrast-enhanced computed tomography (CT) scan of the neck showed a large, hypervascular soft tissue mass with splaying of the left internal carotid artery. Intraoperatively, the tumor was found to be arising from the vagus nerve. Macroscopic surgical pathology examination showed a tan-red, ovoid, and firm mass. Histopathology showed a benign spindle cell tumor with Antoni A areas with palisading cell nuclei and some degenerative change, confirming the diagnosis of vagus nerve schwannoma.

Conclusions: Vagus nerve schwannomas should be distinguished from other tumors that arise in the neck before planning surgery, to minimize the risk of nerve injury. Physicians need to be aware of the differential diagnosis of a neck mass, investigations required, the surgical treatment and the potential postoperative complications.

MeSH Keywords: Neurilemmoma • Vagus Nerve Diseases • Vocal Cord Paralysis

Full-text PDF: <https://www.amjcaserep.com/abstract/index/idArt/904084>

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Background

Schwannoma of the vagus nerve is a benign tumor that usually presents as an asymptomatic slow growing mass, and its occurrence in the neck is rare. The diagnosis may be difficult to make and complete surgical excision is challenging due to the proximity of the vagus nerve from which it originates. Schwannomas usually present as an asymptomatic, well-circumscribed, encapsulated mass growing slowly at a rate of approximately 2.5 mm to 3 mm per year [1].

The most common symptom associated with schwannoma of the vagus nerve arising in the neck is hoarseness due to vocal cord palsy, which can be associated with an involuntary cough during palpation of the mass. Histologically, the tumor is composed of bundles of spindle cells, which are strongly positive for S-100 protein detected using immunohistochemistry [2].

Case Report

We report a case of a 55-year-old woman with a past medical history of hypertension who presented to the clinic complaining of throat irritation and feeling of something stuck in her throat for the past three months. On examination, a bulging left parapharyngeal mass was noted displacing the left tonsil and uvula medially and associated with left neck fullness. A contrast-enhanced computed tomography (CT) scan of the neck showed a large, hypervascular soft tissue mass with splaying of the left internal carotid artery. Extensive varices were noted to be present throughout the face and neck. The left internal jugular vein was partially occluded at the level of and inferior to the mass, possibly secondary to compression by the mass (Figure 1).

During surgical excision, the tumor was found to be arising from the vagus nerve. On cut section, the tumor had a soft center that was suggestive of either cystic or necrotic change. The approximate size of the mass was 4.7×3.6×3.0 cm. The origin appeared to be at the level of the left carotid artery bifurcation.

Due to the size of this parapharyngeal space tumor, a decision was made to resect it surgically, after weighing the risks and benefits of the procedure with the patient. Intraoperatively, the tumor was found to be arising from the vagus nerve. During the resection of the tumor, all attempts were made to peel the outer capsule off the tumor to preserve the nerve branches. The central portion of the tumor was moderately firm and was removed in one piece.

Macroscopic pathology examination showed an ovoid, firm, tan-red tumor that was not well-encapsulated, measuring 4.5×3.5×2.7 cm. Two firm nodules were identified measuring

Figure 1. Contrast-enhanced computed tomography (CT) scan of the neck showing a hypervascular soft tissue mass.

Figure 2. Photomicrograph of the histology of the tumor showing moderate and sparse cellularity of a spindle cell tumor. Hematoxylin and eosin (H&E) (×100).

1.5×0.8×0.6 cm and 1.8×1.5×0.7 cm. Histopathology showed two main histological patterns, consistent with a diagnosis of benign schwannoma. Less cellular areas (Antoni B) contained edematous stroma in which fibers and cells form no distinctive pattern (Figure 2), More cellular areas (Antoni A) included palisading of cell nuclei of spindle and round cells (Verocay bodies) (Figure 3). There were also spindle cells with degenerative atypia (Figure 4). These histological features were consistent with a diagnosis of benign schwannoma of the left vagal

Figure 3. Photomicrograph of the histology of the tumor showing Verocay bodies consisting of palisading cell nuclei, and more cellular Antoni A areas. Hematoxylin and eosin (H&E) (×100).

Figure 4. Photomicrograph of the histology of the tumor showing spindle cells with some degenerative ('ancient') change, and some cellular atypia in keeping with reactive change Hematoxylin and eosin (H&E) (×400).

nerve associated with degenerative changes. The two other nodules were benign, reactive lymph nodules.

Postoperatively, the patient developed left vocal cord paralysis, for which she underwent left medialization thyroplasty. However, the hoarseness of her voice persisted following surgery.

Discussion

Neurogenic tumors of the parapharyngeal space are rare, with 55% of these tumors being schwannomas and approximately

half of these arising from the vagus nerve [3]. Most schwannomas occur between the third and sixth decade of life, affecting both genders equally, and have minimal risk of malignant transformation. Schwannoma presents as a slow-growing, fixed, and painless mass. When schwannoma arises in the neck, hoarseness of voice is the most common symptom. However, as in this case, a pathognomonic clinical sign is a cough on palpation of the mass [4].

Schwannoma of the vagus nerve should be differentiated from other tumors in the neck before planning surgery to minimize the risk of nerve injury due to proximity of the tumor to the vagus nerve. Fine-needle aspiration cytology (FNAC), ultrasonography, computed tomography (CT), and magnetic resonance imaging (MRI) are all preoperative diagnostic methods, although MRI appears to be the investigation of choice for the diagnosis and identification of the nerve of origin.

In this case, the tumor showed two main histological patterns, consistent with a diagnosis of benign schwannoma. The more cellular areas (Antoni A) include palisading of cell nuclei of the spindle cells and round cells (Verocay bodies); the less cellular areas (Antoni B) contain edematous stroma in which fibers and cells form no distinctive pattern [3]. Using immunohistochemistry, schwannoma typically shows bundles of spindle cells, which are strongly positive for S-100 protein [2].

Surgical excision is the treatment of choice for vagal schwannoma and the surgical approach taken will depend on the tumor size, location, and proximity to the vagus nerve and vessels of the neck. The main postoperative complication is hoarseness of the voice due to vocal cord palsy, sometimes associated with coughing and choking while eating [4]. This patient developed left vocal cord palsy after surgery, and despite undergoing left medialization thyroplasty, her voice was permanently affected.

Conclusions

Schwannoma arising from the vagus nerve in the neck is a rare tumor that is usually asymptomatic but requires surgical excision. Physicians need to be aware of the diagnostic work-up and differential diagnosis of a neck mass, the surgical treatment, and the postoperative complications, and preoperative planning is also important. The prognosis of schwannomas after complete excision is good, and recurrence is rare.

Competing interests

The authors declare that they have no competing interest.

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