Vagus nerve schwannoma, an uncommon benign neoplasm: case report

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Abstract

Schwannomas are rare benign tumors that originate in the nerve sheath. They occur 25 to 45% of the time in the head and/or neck, with a low risk of malignancy. The symptoms are nonspecific, which makes diagnosis challenging, and surgery is the only therapeutic option. Given the rarity of this pathology, the focus of this report is to present the case of a young patient for whom infectious or malignant etiologies causing cervical masses were ruled out, leading to a diagnosis of vagus nerve schwannoma. This tumor should be considered in the differential diagnosis.

Keywords: neurilemmoma; head and neck neoplasms; head and neck surgery; case reports.

Introduction

Schwannomas, also known as neurilemomas, are rare benign tumors originating from the Schwann cells that form the myelin sheath. These tumors can affect any nerve, though they infrequently present in the head and/or neck region, typically manifesting in a parapharyngeal location and a vagal origin. Conversion to malignancy is rare, with a prevalence of 4% in cervical tumors.

Case report

A 30-year-old male, an occasional smoker with a history of toxoplasmosis and rubella during childhood, and genital HPV diagnosed 18 months prior, presented for medical evaluation because of a left cervical mass that had been present for six months, without accompanying symptoms. Physical examination revealed lymphadenopathy in right level III and a painless mass of cystic consistency in the left cervical region, approximately 4 centimeters in diameter, between levels III and IV (Figure 1).

Oroscopy and laryngoscopy showed normal macroscopic findings. Laboratory tests indicated no inflammatory response, and the thyroid profile was normal. Infectious diseases were ruled out. Ultrasound revealed oval nodes of 7.2 and 6.5 mm in right level III, with well-defined edges and a good hilum-cortex...
relationship, appearing inflammatory. In the left level IV, mixed nodulation was observed, featuring solid and internal liquid areas measuring 47 x 30 x 27 mm, with a volume of 20 milliliters, and displaying peri- and intra-nodular vascularization in the solid areas, hypoechoic, rounded, with well-defined edges, and posterior acoustic reinforcement (Figure 2).

**Figure 1.** 30-year-old man with a left cervical mass between levels III and IV.

**Figure 2.** Ultrasound image of mixed nodulation in the left level IV, displaying internal solid and liquid areas, measuring 47 x 30 x 27 mm with a volume of 20 milliliters.
To rule out malignancy, cytology was performed, which indicated an epithelial lesion not conclusive for malignancy; therefore, a core biopsy was performed. Histological sections showed fibrous tissue, nerve fillets, and lymph node parenchyma with small lymphocytes. Surgery was decided upon to excise the left cystic lesion and perform a frozen biopsy of the right lymphadenopathy, both testing negative for malignancy. During surgery, it was determined that the cystic lesion was dependent on the vagus nerve, hence diagnosed as a schwannoma. Partial resection was performed, including excision of the cystic portion with nerve fiber preservation and marsupialization towards the sternocleidomastoid and thyrohyoid muscles (Figure 3).

Figure 3. Cystic lesion dependent on the left vagus nerve.

Post-surgery, the patient was in good condition with no neurological deficit and was discharged after 48 hours. He attended a medical check-up one year post-surgery without disease recurrence, maintaining adequate voice tone and without any associated symptoms or deficits.

The authors declare that written consent for publication was obtained from the patient.

Discussion

Schwannomas, or neurinomas, are benign encapsulated neoplasms formed by Schwann cells. The typical age of presentation ranges between the second
and fifth decades of life, with no predilection for race or sex. While many neurinomas are identified sporadically, they can also be associated with neurofibromatosis type 2 or the Carney complex. The most common symptom is a solitary mass, accompanied by nonspecific signs and symptoms that vary depending on the nerve of origin, location, extent, and involvement of adjacent structures. This can cause dysphagia, nasal obstruction, dyspnea, or hoarseness. Neural deficit occurs in large tumors or when there is neural compression, such as in intracranial skull base tumors.

Diagnosing schwannomas is challenging because the symptoms are nonspecific. Cytology is often inconclusive but useful in ruling out malignancy, as in our case where the patient had an asymptomatic cervical mass. Complementary studies yielded negative results for infectious and oncological diseases. Histopathological examination revealed a biphasic histological pattern with areas of high cellularity containing spindle cells organized in bundles or eddies with a group of parallel palisaded nuclei known as Verocay bodies in the Antoni B areas. Cells characteristic of cystic degeneration or xanthomatous alterations were observed in the Antoni A areas, and immunohistochemistry was positive for S-100 protein. Imaging examinations typically show a cystic lesion, sometimes associated in with necrosis, which contributes to planning surgery by providing data on size, extent, and location. Magnetic resonance imaging specifically can show dispersed flow similar to “salt and pepper,” which guides diagnosis.

Surgical removal is the only treatment option; techniques include resection of the tumor with primary anastomosis of the nerve, using a neural graft, enucleation of the tumor among healthy nerve fibers, or emptying of the tumor while preserving the capsule, though the last presents a high risk of recurrence. Because of the generally benign nature of this pathology, efforts should focus on preserving nerve functionality to ensure a good quality of life. In this case, partial resection of the cyst with nerve fiber preservation and marsupialization towards adjacent cervical muscles was performed, yielding good post-surgical outcomes without neurological deficits or recurrence. In cases of suspected malignancy, enucleation or partial excision is not recommended. The technique of performing an enucleation between the nerve fibers is based on the fact that the tumor’s capsule layer is composed of epineurium and perineurium, which are longitudinally incised to expose the true capsule, enabling cyst removal without compromising the nerve. This approach preserves nerve function by more than 30% compared to tumor resection and primary anastomosis. A nerve stimulator may be used during surgery to ensure accurate identification of nerve fibers, minimizing the risk of their resection. The recurrence rate for enucleation is similar to that for resection and primary anastomosis. The most common post-surgical complication is an altered voice tone with hoarseness; the prevalence of post-surgical vocal cord paralysis is approximately 12%.1,4,5

References


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