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Nerve**

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CASE REPORT

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CT Features of an Unusual Calcified Schwannoma of the Superior Laryngeal Nerve

SUMMARY: We report the CT findings in a patient with a lateral neck mass histologically diagnosed as a laryngeal schwannoma but presenting some uncommon CT features. CT showed unusual calcified components, very rarely observed and potentially misleading for diagnosis. However, this imaging feature can be found in ancient schwannomas. Our case is, therefore, a very rare one and reviews the main differential diagnoses.

Schwannomas are benign tumors arising from Schwann cells surrounding peripheral and cranial nerves. These neoplasms account for approximately 5% of all benign soft-tissue tumors.¹ All ages are affected, but there is a peak incidence between the 4th and 6th decade. Generally, there is no sex-ratio difference. Although most schwannomas are sporadic, there is a high incidence of schwannomas in patients with type 2 neurofibromatosis.²

Calcification in schwannoma is extremely rare and may cause difficulty in establishing a neuroradiologic differential diagnosis. We present the CT findings of an unusual calcified laryngeal schwannoma and review the main differential diagnoses.

Case Report

Clinical History and Physical Examination Findings

A 37-year-old man with no clinical history presented to an ear, nose, and throat specialist with a 6-year history of a slowly enlarging mass in the lateral left neck, recently impairing his swallowing. The evaluation revealed a 3-cm palpable, nonmobile when swallowing, nontender nonpainful lateral cervical mass. Examination of the oral cavity revealed no clinical sign involving the floor of the mouth. There were no pathologic lymph nodes in the neck. The findings of a fiber optic nasopharyngeal scope examination revealed normal movement of the vocal cords. The remainder of the examination yielded no other pathologic findings.

Radiographic Findings

Ultrasonography showed a calcified vascularized cervical mass. A contrast-enhanced CT scan of the neck (Fig 1) revealed a heterogeneous-attenuation lesion with well-defined margins, located in the submandibular space. Most of the mass appeared hypoattenuated, with some mild contrast-enhanced focal areas and a large calcification in the middle of the mass. There was no infiltration of the surrounding fat. The mass had deformed the hyoid bone left greater cornua, typical of a slow-growing benign chronic lesion. There were no prominent lymph nodes in the cervical group.

Histologic Findings

The surgeon performed a total resection of an encapsulated mass arising from the left superior laryngeal nerve. The tumor was mostly hypocellular, with myxoid fibrotic stroma, thick hyaline wall vessels, and dystrophic calcifications. Verocay bodies and S-100 protein were present. No mitoses, atypia, or areas of necrosis were identified. The histologic diagnosis of the tumor was consistent with an ancient schwannoma.

Discussion

Schwannomas are benign slow-growing tumors that arise from the Schwann cells of any peripheral, cranial, or autonomic nerve sheaths in various anatomic locations. Schwannomas involving the head and neck are most commonly intracranial and usually involve the vestibular nerves. Between 25% and 45% of extracranial schwannomas occur in the head and neck region.³ The tumor can develop anywhere from the base of the skull to the thoracic inlet but is most commonly found in the mid neck. Schwannomas in the neck can arise from spinal or cranial nerve roots. Most tumors arise from the glossopharyngeal, vagus, accessory, and hypoglossal nerves; and sympathetic chains are located in the medial aspect of the neck in the compartment of the parapharyngeal space. In the lateral aspect of the neck, most tumors arise from the cutaneous or muscular branches of the cervical or brachial plexus.^{3,4}

The occurrence of schwannomas in the larynx is very rarely reported as an unusual cause of dysphagia, dyspnea, and dysphonia, revealing the tumor.⁵⁻¹⁰ These tumors mainly involve the aryepiglottic folds or the ventricular folds, or they may extend to the pyriform fossa and, more rarely, to the subglottic area. However, in our case, the schwannoma was not located within the larynx, even though the superior laryngeal nerve was involved.

Schwannomas are usually solitary and typically manifest as a slowly enlarging painless mass. Pain and neurologic deficit are uncommon but suggestive of malignancy.^{3,11} CT or MR imaging findings of schwannomas are often similar to those of neurofibromas and, in many cases, the 2 cannot be distinguished. Usually, the lesion is a large sharply demarcated mass, round or oval, isoattenuated with muscle, sometimes cystic, and often heterogeneously enhancing.¹²

Calcified schwannomas in the head and neck are extremely rare. However, this imaging feature can be found in ancient schwannomas. They are a rare variant of schwannoma characterized by degenerative changes resulting from the long-term progression of the tumor. Calcification is a usual degenerative change; however, it is rarely detected on radiologic

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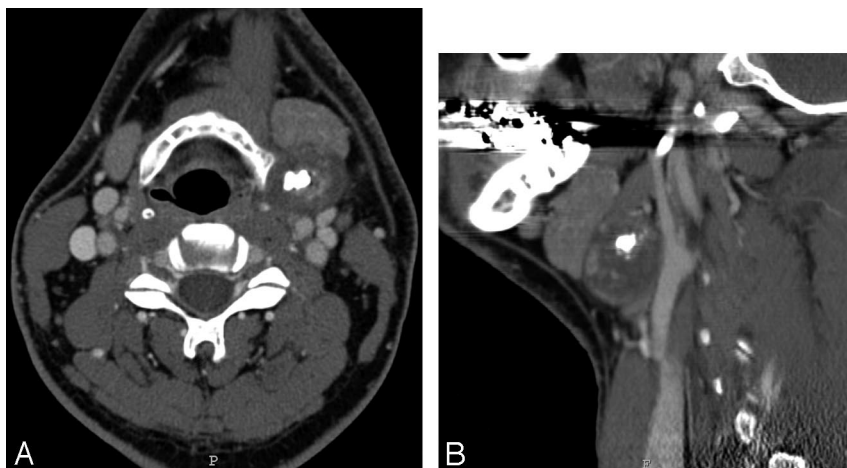


Fig 1. Contrast-enhanced CT of the neck shows a 3-cm heterogeneous tumor located between the submandibular gland and the carotid space, with a single large calcification. Note the mass effect on the left greater cornua of the hyoid bone, typical of a slow-growing benign tumor. A, Axial view. B, Sagittal view.

examination.¹³ Only a few reports described cases of ancient schwannomas located in the head and neck region. In these reports, all the tumors were benign, with the exception of 1 malignant schwannoma.¹⁴ However when observed, calcifications are usually scattered, whereas we encountered a single large one.

Schwannomas with these degenerative changes can be misdiagnosed as other forms of soft-tissue neoplasms. In our case, the schwannoma was located in the anterior anatomic triangle of the neck based on the division of the sternocleidomastoid muscle. The imaging approach based on spaces located the tumor at the level of the hyoid bone, at the posterior aspect of the submandibular space, anterior to the carotid space, with an inferior extension to the anterior cervical space. Therefore, a differential diagnosis should include congenital mass, such as a second branchial cleft cyst, and inflammatory processes such as an abscess; or adenopathy, diving ranula, and especially benign tumors such as epidermoid or dermoid cysts; and the tail of a parotid pedunculated tumor. However, whereas a cystic schwannoma can mimic a 2nd branchial cleft cyst,¹⁵ the latter diagnosis would have been most unlikely in our case because of its heterogeneous aspect, unless the cyst had been previously infected. Moreover the large calcification excluded the diagnosis of second branchial cleft cyst. This calcification in our patient was very suggestive of dermoid cyst or granulomatous adenopathy, especially tuberculous adenitis. A submandibular tumor was excluded because the gland was anteriorly displaced and not infiltrated. As for neurofibroma, clinical and pathologic features usually allow a sure diagnosis.

The definitive diagnosis remains histologic. Pathologically, schwannomas are usually encapsulated tumors. The tumor is composed of spindle-shaped neoplastic Schwann cells with alternating areas of compact elongated cells (Antoni A) and less cellular areas (Antoni B).² Verocay bodies and immunopositive S-100 protein are typical. Prominent vessels with thick hyaline walls are usually present. Additional microscopic features seen in schwannomas, especially in ancient ones, include occasional foci of necrosis, cystic degeneration, foam cells, hemosiderin, calcification, and attenuated bands of hyalinized connective tissue.¹⁶

In conclusion, to our knowledge, this is the first report of a calcified schwannoma of the superior laryngeal nerve. Both location and single calcification were very unusual. Although rare, schwannoma becoming ancient should be included in the differential diagnosis when a calcified lateral cervical mass is identified.

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