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Tumor Discovery

CASE REPORT

Calcified peripheral schwannoma mimicking a cervical lymph node: A case report and literature review

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Abstract

Peripheral schwannomas are typically benign tumors originating from Schwann cells of peripheral nerves. Although often asymptomatic, such tumors may present diagnostic challenges since they resemble other known structures, leading to potential misinterpretations. Here, we present the case of a 17-year-old patient with a cervical mass that initially posed a diagnostic dilemma, resembling a cervical lymph node on clinical examination. The calcified nature of the tumor added complexity to its identification, and our case underscores the significance of a good diagnostic evaluation. The literature on calcified peripheral schwannomas is scarce, but the current case contributes to the understanding of atypical presentations. Diagnostic modalities, including imaging studies and histopathological examination, play a pivotal role in confirming the schwannoma diagnosis and differentiating it from other entities. Surgical excision remains the primary treatment, but awareness of varied presentations is crucial for accurate management. This case report serves as a valuable addition to the literature, highlighting calcified peripheral schwannomas mimicking cervical lymph nodes. Medical practitioners should maintain a high index of suspicion for schwannomas in unusual clinical cases, ensuring accurate diagnosis and appropriate management.

Keywords: Cervical mass; Schwannoma; Calcification

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1. Introduction

Cervical schwannomas are benign tumors of the peripheral nerves developed exclusively from Schwann cells. Initially described by Verocay in 1908,[1] they are estimated to be present in the neck region in 25 % of all cases.[2] The nerve of origin remains unidentified in 50% of the cases,[3] but they are found to be predominantly formed within the vagus nerve.[2] Other structures such as the sympathetic cervical chain can be, although rarely, the origin of this benign entity.[2] Here, we report the case of a chronic cervical schwannoma, which was initially misinterpreted as a cervical lymph node.

2. Case presentation

The present case revolves around a 17-year-old girl with no significant medical history, who reported a 2-year history of progressive swelling in the upper right laterocervical

region behind the mandibular angle. The patient exhibited no signs of numbness, compression (such as dysphagia or dyspnea), or discomfort. On examination, a firm painless mass, approximately 2 cm × 2 cm, was palpable at level 2 on the right side. The mass was mobile in both superficial and deep planes, with no skin changes overlying. Oto-rhino-laryngoscopic examination revealed normal findings. Ultrasound examination identified a well-defined solid cystic mass, measuring 2.5 cm in diameter, hypoechoic with no internal flow. There was no continuity between the proximal and the distal ends. Infracentimetric lymph node structures were identified alongside the mass. Given their proximity, the original diagnosis of pathological lymph node was maintained, and thus, no further radiological exploration was pursued. A fine-needle aspiration cytology (FNAC) of the mass showed an atypia of undetermined significance. Surgical exploration under general anesthesia revealed a white nodular formation located medially to the anterior border of the sternocleidomastoid muscle. The mass was distant from the neurovascular bundle and easily dissected from the surrounding structures. A complete excision was carried out (Figures 1 and 2).

Histologically, the nodular mass was calcified, exhibiting necrotic rearrangement. Histopathologic examination revealed spindle-shaped and elongated tumor cells, which showed no mitotic activity, pointing to a plausible diagnosis of schwannoma (Figure 3). The post-operative course was not marked with any remarkable clinical changes, and after a one-year follow-up, the patient showed no signs of recurrence.

3. Discussion

Schwannomas are benign tumors that originate in any peripheral nerves, with the exception of the olfactory and optic nerves.[4] They affect the head-and-neck region in 20 – 45% of cases, with the vestibular nerve being the primary origin of development.[5] The occurrence of schwannomas is common in patients between 20 and 50 years old, regardless of gender.[4] The clinical signs of cervical schwannomas depend on various factors including the location, size, and the nerve of origin. The most typical clinical presentation is isolated laterocervical mass, which is asymptomatic and characterized by slow mass growth.[5] Additional symptoms may be present in some cases, which can be explained by an external mass compression of the oro-pharyngolaryngeal axes or the adjacent nerves.[4] In our case, the patient did not exhibit any symptoms other than progressive swelling.

Identifying the nerve of origin in cervical schwannomas poses a significant challenge. This is evidenced by a comprehensive case study involving patients with non-vestibular schwannomas of the head and neck, where specific nerve of origin could only be determined in 16 out of 26 patients (62%), with the origin of the remaining cases, presumably from unnamed small nerve plexus branches, left unidentified.[4] Similarly, an article from India highlights the difficulty of determining the nerve of origin for this tumor entity, suggesting a successful chance of identification as low as 50%.[3]



Figure 1. Peroperative image showing the nodular mass.



Figure 2. Excised mass before histological examination.

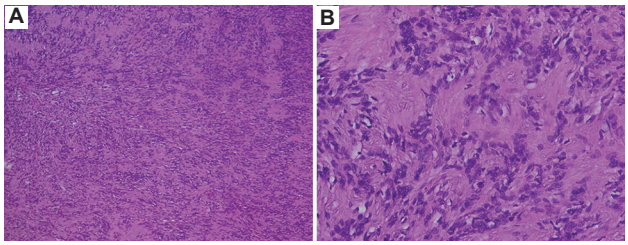


Figure 3. Microscopic examination of hematoxylin-eosin-stained sections of surgically resected tumor under ×20 (A) and ×40 (B) magnification. The sections show cytologically bland spindle cells against a vague nuclear palisading and fibrillary background.

Whether surgical approach is employed to resect cervical schwannomas is determined by the location and size of the mass. For instance, accurately identifying location of the mass on vital structures such as neurovascular bundle and spinal nerve is crucial for decision-making on whether a horizontal or Paul André incision should be employed.[3,6] In our case, the nerve of origin was not identified. However, considering the location of the mass and the ease with which it was dissected from the surrounding structures during the surgical exploration, we also believe that the origin was likely a small nerve of the plexus branches.

Although the definitive diagnosis relies strictly on histopathologic means, pre-operative imaging proves to be an essential tool for diagnosing cervical schwannomas. Imaging can yield crucial information regarding the size and location of the mass, as well as the involvement of surrounding structures, such as the neurovascular bundle, and provide guidance in performing a fine need aspiration cytology or biopsy (FNAC or FNAB, respectively) through ultrasonography.[4,7] Both computed tomography (CT) and magnetic resonance imaging (MRI) findings assist with ruling out differential diagnoses such as paraganglioma or congenital cysts. These radiological modalities can also help identify the nerve of origin by analyzing the mass effect on the vascular axis.[6] In the case of the involvement of vagus nerve, the tumor tends to widen the space between the internal carotid or common carotid artery and the internal jugular vein, whereas the involvement of sympathetic nerve displaces the jugulocarotid axis forward.[8] Ultrasonographic findings alone can be insufficient to differentiate between schwannomas and pathologic lymph nodes, especially since the exploration is operator dependent.[9] In a comparative study, Ahn *et al.* reported that core needle biopsy (CNB) delivered a high-accuracy performance in diagnosing extracranial schwannomas of the head and neck (96.5% specificity and 100% sensitivity), as compared to FNAC.[7] However, despite its relatively low sensitivity in this respect,[10] FNAC can be used to rule out other potential malignant lesions.[9] A FNAC was performed in our case guided by ultrasonography showing an atypia of undetermined significance.

While the tumor progression is typically slow, and malignant transformation is exceptionally rare, complete excision of the lesion remains the optimal treatment for symptomatic cervical schwannomas.[5,6] Claude-Bernard-Horner syndrome (miosis, ptosis, anhidrosis, and enophthalmos) is the most common post-operative complication in schwannomas of the cervical sympathetic chain.[6] This complication can be confirmed in an anatomopathological examination by identifying spindle cells with Verocay bodies or without tissue cellularity.[11] In addition to their morphological characteristics, schwannomas can be differentiated from leiomyomas and palisaded myofibroblastoma by virtue of their distinctive immunoreactivity profile.[9] Schwannomas typically demonstrate diffuse expression of S100 protein, while showing no reactivity for antibody against muscle markers such as desmin and smooth muscle actin.[12]

Calcification is a rare occurrence in schwannomas. This is supported by a study by Din *et al.* who identified only 27 cases of calcifications out of 2116 cases of schwannoma, of which only three cases were detected in the cervical region.[13] Recurrence of schwannoma is exceedingly uncommon, even in cases where the tumor is incompletely resected.[14]

4. Conclusion

Schwannoma should be considered a differential diagnosis in patients with a neck mass due to the potential misinterpretation as lymphadenopathy, even if ultrasonography screening is performed. Radiological examinations including CT scan and MRI play an important role in accurate diagnosis-making as well as pre-surgical analysis of anatomical structures.

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Conflict of interest

The authors declare that they have no competing interests.

Author contributions

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Ethics approval and consent to participate

Written consent was obtained from the patient and his family to participate in this study.

Consent for publication

Written consent was obtained from the patient and his family for the publication of this case report and accompanying images.

Availability of data

Not applicable.

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