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Silent Growth: 18-Year History of an Upper Lip Schwannoma

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Abstract

Schwannomas are benign, encapsulated tumors originating from Schwann cells, commonly found in the head and neck region. Although rare, lip schwannomas can occur and are typically asymptomatic. This case report describes an 18-year history of a slow-growing upper lip schwannoma in an 18-year-old patient. The tumor, which developed after a traumatic injury, presented as a painless, firm mass without pulsation and was diagnosed through histopathological and immunohistochemical analysis. The tumor was surgically excised, and follow-up revealed no recurrence. The prognosis is generally favorable with complete surgical removal, and regular follow-up is advised to monitor for recurrence.

Keywords

Schwannoma, Upper Lip, Trauma, Histopathology

1. Introduction

Schwannoma is a benign tumor that arises from Schwann cells, which form the myelin sheath around myelinated nerve fibers. While schwannomas are typically solitary, they can also present as multiple tumors, especially in association with neurofibromatosis. Although most schwannomas are benign, approximately 2% are reported to undergo malignant transformation, potentially leading to distant metastasis (Hilton & Hanemann, 2014). Schwannomas of the oral cavity are rare. They have been reported in areas such as the tongue, palate, and buccal mucosa. Lip schwannomas, however, are exceptionally uncommon, with only a handful of cases documented in the medical literature.

We report a rare and intriguing case of schwannoma in the lower lip of a 21-

year-old patient, developed over 18 years following a traumatic fall, which made the diagnosis particularly unexpected. The tumor was painless, firm, and slow growing. The diagnosis was confirmed through histopathological analysis, and the lesion was successfully treated with surgical excision. This case highlights the importance of considering schwannomas in the differential diagnosis of lip masses such as traumatic fibroma, labial minor salivary gland tumor, or trauma-related swelling. It represents one of the very few documented instances of schwannoma in the lip and is likely only the second case reported in association with a traumatic history (Desai, 2019). This highlights the potential role of trauma as a contributing factor in the development of such lesions.

2. Case Report

A 21-year-old patient, a nonsmoker with no other significant personal medical history, there were neither associated medical findings nor family history about any other concomitant diseases such as neurofibromatosis. He visited the military hospital with a complaint of painless swelling on the left upper lip. The swelling first appeared 18 years earlier following a traumatic injury and gradually increased in size over time. Upon examination, the swelling was localized on the left side of the upper lip (**Figure 1**). It was a firm mass measuring 30 mm × 20 mm, with normal-colored and well-contoured overlying mucosa, painless, and no pulsation. No other medical findings or family history of conditions such as neurofibromatosis were reported.

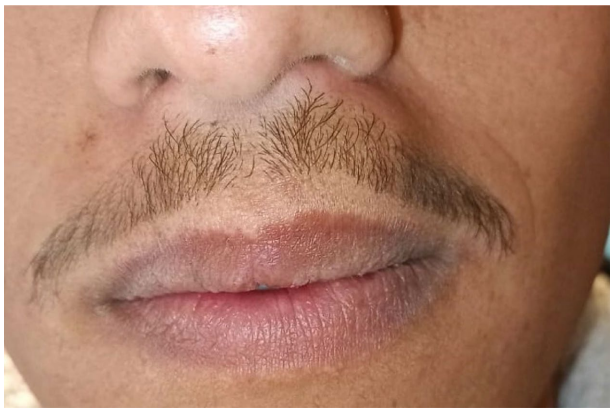

Figure 1. Extraoral examination revealed swelling of the upper lip.

Based on the patient's history and clinical assessment, the differential diagnoses included traumatic fibroma, labial minor salivary gland tumor, or trauma-related swelling. The ultrasound scan revealed a well-limited, homogeneous, isoechoic, non-vascular mass. The lesion was treated with complete surgical excision under local anesthesia, followed by primary closure (**Figure 2**). Its encapsulated nature allowed for precise and total removal. The patient was discharged with routine analgesics and antibiotics. The excised specimen was sent for histopathological analysis.

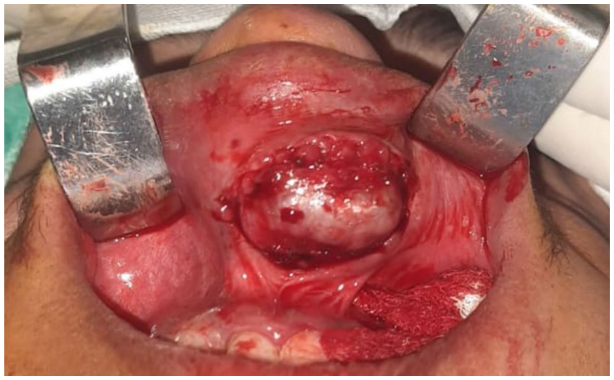

Figure 2. Intraoperative image showing excision and evacuation of the tumor. And their macroscopic appearance.

Histopathological evaluations confirmed the diagnosis of schwannoma, showing a well-limited, encapsulated fusocellular proliferation with a storiform organization and a biphasic appearance, with cellular and poorly cellular zones. The cells are spindle-shaped with wavy nuclei, the site of rare mitosis. The cytoplasm is fibrillar and abandoning. The stroma is fibrous, harbouring thin-walled blood vessels.

Clinical monitoring at the 7th and 15th day follow-up showed excellent healing with no wound dehiscence, infection, or residual swelling in the surgical area (**Figure 3**) and all mucosal and lip sutures were successfully removed.

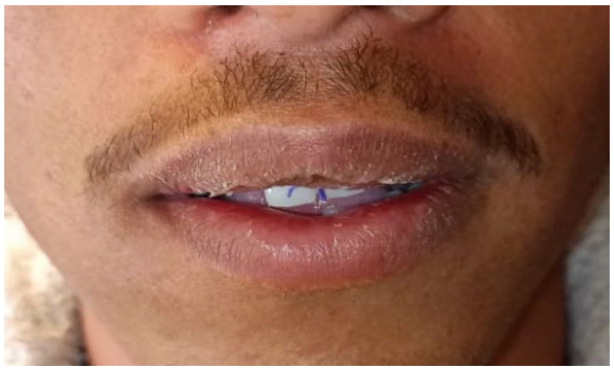

Figure 3. Evaluation on the 15th day postoperative showed no dehiscence of the wound, no signs of infection, and no swelling.

3. Discussion

Schwannoma is a benign, encapsulated, slow-growing and generally solitary tumor that arises from Schwann cells of the peripheral nerve sheath, which can often undergo cystic and degenerative change. They usually arise from peripheral nerves, especially on the deep parts of the soft tissues (the acoustic nerve, posterior spinal root, and the extremities, trunk, body, and neck) (Kara & Topuz, 2002). In the head and neck region, the tumor can occur either in peripheral cranial nerves or intracranially (acoustic nerves are the most common intracranial location). In

the head and neck region, the most involved nerves are the vagus and the cervical sympathetic chain (Saydam et al., 2000).

Approximately 25% - 45% of all schwannomas are seen in the head and neck region. Of these, approximately 1% - 12% occur intraorally with a predilection for the tongue. The palate, buccal mucosa, lips, and gingiva are also affected in decreasing order (Harazono et al., 2022). According to Rizqan Maulana et al., only 13 cases of lip schwannomas have been reported in the literature (Maulana et al., 2024). The highest incidence occurs in the third and fourth decades of life, more rarely observed at a younger age, and the prevalence is not significantly different between males and females (Desai, 2019). This case reports a rare localization of schwannoma affecting the lower lip, occurring at a younger age.

Many schwannomas are asymptomatic, but when symptoms do occur, swelling is the most common presenting feature, followed by paraesthesia. In cases where the tumor is located in the intraosseous regions of the mandible, it may cause pain or sensory disturbances. These symptoms typically result from local compression or bony erosion (Salehinejad et al., 2017).

The tumor is usually characterized by a solitary, slow-growing mass with a smooth surface, whose structure can vary from fluctuating cyst to solid. Likewise, in our case, the tumour was solid and slow growing.

According to a retrospective analysis of schwannoma in the oral and maxillofacial region, the average tumor size was 13.2 mm, with a range of 2 to 53 mm (Harazono et al., 2022). In our case, the tumor size was 30 mm × 20 mm.

Asaumi et al. (2000) reported characteristic features of schwannomas of the upper lip on ultrasonographic and advanced imaging (computed tomography and magnetic resonance imaging) methods. On ultrasonographic imaging, homogenous and hypoechoic findings and posterior acoustic enhancement were reported. On computed tomography imaging, it has been reported as a marginated mass with homogenous soft-tissue density. Unlike these findings, on magnetic resonance imaging, it has been defined as a homogenous lesion with low-intermediate signal intensity on T1-weighted and high signal intensity on T2-weighted images. However, the authors emphasized that radiologic imaging should not be considered routine or necessary for differential diagnosis, particularly due to the small size of these tumors in the lip. In this case, we did not perform any radiologic imaging (Bayindir et al., 2013).

Schwannoma is categorized into seven histopathological subtypes: classic (Verocay), cellular, plexiform, cranial nerve, melanotic, degenerated, and granular cell Schwannoma. Typical histological features include the presence of Antoni A areas, Antoni B areas, or both. Antoni A regions are hypercellular and consist of densely packed spindle-shaped Schwann cells arranged in palisades, while Antoni B regions are hypocellular, featuring small round cells within a myxoid stroma. The Verocay body, a hallmark feature, is formed by parallel rows of nuclei separated by hyalinized collagen bands. Notably, these lesions typically exhibit low vascularity, with rare occurrences of necrosis or mitotic activity (Dos Santos et al.,

2011; Melzer et al., 2017).

Schwannomas can develop either spontaneously or as part of familial tumor syndromes like neurofibromatosis type 2 (NF2), schwannomatosis, or Carney's complex (Hilton & Hanemann, 2014), or post-radiation. Traumatic origin is not frequent.

Syndromes like neurofibromatosis type 2 is an autosomal dominant syndrome characterized by the development of multiple schwannomas. The diagnosis of NF2 is based on clinical evaluation using the established Manchester criteria for neurofibromatosis type 2. A comprehensive diagnostic approach includes routine clinical assessment, detailed family history, a thorough cutaneous examination, slit-lamp ophthalmic evaluation, craniospinal magnetic resonance imaging (MRI), and molecular genetic analysis (Evans, 2009).

Schwannomatosis is characterized by multiple painful schwannomas, typically occurring in the neck, trunk, and extremities, while visceral involvement is rare, with most cases being sporadic and only 15% - 25% having a family history. Unlike NF2, vestibular schwannomas are rare, and diagnostic criteria (**Table 1**) include molecular findings to distinguish them from NF2 (Plotkin et al., 2013).

Table 1. Manchester criteria for diagnosis of neurofibromatosis type 2 (Hilton & Hanemann, 2014).

1) Bilateral vestibular schwannoma (VS) or family history of NF2 plus: Unilateral VS or two of meningioma, glioma, neurofibroma, schwannoma, posterior subcapsular lenticular opacities.
2) Unilateral VS plus two of meningioma, glioma, neurofibroma, schwannoma, posterior subcapsular lenticular opacities.
3) Two or more meningioma plus unilateral VS or two of glioma, neurofibroma, schwannoma, cataracts.

NF2 = neurofibromatosis type.

Carney's complex is characterized by a combination of features, including conjunctival and mucosal lentigines, myxomas (particularly cardiac), schwannomas, and various endocrine abnormalities such as acromegaly, Sertoli cell tumors, and Cushing's syndrome (Wilkes et al., 2005). Diagnostic criteria (**Table 2**) have been established to aid in its identification (Stratakis et al., 2001).

The prognosis for schwannoma is generally favorable. The preferred treatment is conservative surgical removal, as wide excision is not recommended. When complete enucleation is achieved, recurrence is unlikely. The malignant transformation of benign schwannomas remains a topic of debate. Approximately 2% of Schwannomas are reported to be malignant with distant metastases; while some authors question its occurrence, a few isolated cases have been reported (Stratakis et al., 2001). In our patient's case, malignant transformation was not considered, as examination of the excised mass revealed benign microscopic features and confirmed complete removal. Periodic follow-up is very important for early detection of recurrence.

Table 2. Diagnostic criteria for schwannomatosis (Hilton & Hanemann, 2014).

Molecular diagnosis
1) Two or more schwannomas or meningiomas[1] and genetic studies of at least two tumors showing loss of heterozygosity at chromosome 22 and NF2 mutations. The presence of a common SMARCB1 mutation defines SMARCB1-associated schwannomatosis.
2) One schwannoma or meningioma[1] and a germ-line pathogenic SMARCB1 mutation.
Clinical diagnosis
1) Two or more non-intradermal schwannomas (one with pathological confirmation) and the absence of vestibular schwannoma on thin-sliced MRI[2].
2) One schwannoma or meningioma[1] and affected first-degree relative.
3) Possible diagnosis of two or more non-intradermal schwannomas (without pathological confirmation) and chronic pain associated with tumors.
Exclusion criteria: germ-line pathogenic NF2 mutation, fulfill criteria for NF2, first-degree relative with NF2, schwannomas in radiation field only.

[1]Pathologically confirmed. [2]These criteria may include some mosaic NF2 patients, and some schwannomatosis patients may have unilateral vestibular schwannomas or multiple meningiomas. MRI = magnetic resonance imaging; NF2 = neurofibromatosis type 2.

4. Conclusion

Schwannomas are benign, slow-growing tumors that rarely occur in the lip, making this case an unusual and noteworthy presentation. The diagnosis was confirmed through histopathological analysis and surgical excision—the preferred treatment—was successfully performed, offering a favorable prognosis with low recurrence rates when complete removal is achieved. While trauma is rarely associated with schwannoma development, this case raises the possibility of its role in certain instances. However, it is important to acknowledge the limitations inherent in establishing causality based on a single case report. The rarity of this presentation underscores the need for further research to better understand the potential link between trauma and schwannoma development. Regular follow-up remains essential to monitor for recurrence, particularly in atypical cases. This report contributes to the limited literature on lip schwannomas and emphasizes the importance of accurate diagnosis, timely intervention, and consideration of schwannomas in the differential diagnosis of lip masses.

Ethical Approval

The patient was fully informed about the purpose of the case report, the details that would be included, and how it would be used. They provided their voluntary agreement, ensuring compliance with ethical standards and respect for their privacy and rights.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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