

Ancient Neurilemoma of the Lower Lip Mimicking Malignancy: A case report with Histopathologic Challenge

Dr. Kailash Chandra Dash, Dr Asutosh Das, Dr. Noureen Nahar, Dr Shubhangi Gupta,
Dr Sreepreeti Champatray, Dr. Lipsa Bhuyan

ABSTRACT

Neurilemoma, also known as Schwannoma, is a slow-growing, benign tumor of the nerve sheath that is comparatively rare in the oral cavity. The “Ancient” variant of neurilemomas presents a substantial diagnostic problem, whereas regular neurilemomas exhibit a characteristic biphasic pattern. A 65-year-old woman with a hard, painless lump on his lower lip is the subject of this case report. Significant nuclear atypia and hyperchromasia were found during the initial histopathologic evaluation, which raised the possibility of a spindle cell sarcoma. Nonetheless, a diagnosis of Ancient Neurilemoma was confirmed by the robust S100 positive and lack of mitotic features. This case emphasizes how crucial it is to identify degenerative changes in order to prevent needless, drastic surgery.

defined submucosal nodule that measures about 2.5 cm in diameter. There were no signs of ulceration, paresthesia, or discomfort.[Figure-1a] The differential diagnosis charted out were minor salivary gland tumor, mucocele, or fibroma.

Under local anesthesia, a surgical excision was performed. The tumor was found to be well-encapsulated and “shelled out” easily from the surrounding labial tissues.[Figure 1b]

INTRODUCTION

Schwann cell-derived encapsulated tumors are called neuromas. Only approximately 1% of instances are detected intraorally, whereas between 25% and 45% of all cases occur in the head and neck area. The lower lip is an atypical presentation because the tongue is the most prevalent place [1].

Due to the long-standing nature of the tumor, the “Ancient” variant is characterized by “degenerate” changes—specifically, marked nuclear pleomorphism and hyperchromasia. The histopathologic diagnosis is a crucial stage in patient care since these characteristics are often misconstrued as indicators of other spindle cell malignancies such leiomyosarcoma or malignant peripheral nerve sheath tumor [2].

Case Presentation

Clinical Findings

A 65-year-old woman presented with a gradual, asymptomatic swelling on the lower labial mucosa for about six months. On examination a firm, well-

1. Professor, Dept. of Oral & Maxillofacial Pathology, Kalinga Institute of Dental Sciences, KIIT Deemed to be University, Bhubaneswar, Odisha. kcdash1986@gmail.com. ORCID id: 0000-0001-5346-2856
2. Dr Asutosh Das, MDS. Reader, Department of Oral & Maxillofacial Surgery, Kalinga Institute of Dental Sciences, KIIT Deemed to be university. Email id.: asutoshd@gmail.com. ORCID Id. 0000-0001-8845-0054
3. Dr. Noureen Nahar. Senior lecturer, Oral medicine and oral radiology, Kalinga Institute of Dental Sciences, KIIT Deemed to be University. Email: noureen.nahar@kids.ac.in
4. Dr Shubhangi Gupta. Assistant professor, Department of oral and maxillofacial surgery, I.T.S Dental college, Hospital and research centre, Greater Noida. drshubhangi.dntlgn@its.edu.in. Orcid id: 0000-0001-9487-4742
5. Dr Sreepreeti Champatray. Associate Professor, Department of Oral Pathology & Microbiology, Institute of Dental sciences, SOA Deemed to be University, Bhubaneswar, Odisha. Mail: dchampatray@soa.ac.in
6. Correspondence: Dr. Lipsa Bhuyan, Professor, Department of Oral and Maxillofacial Pathology, Kalinga Institute of Dental Sciences, KIIT Deemed to be University, Patia, Bhubaneswar-751024. bhuyanlipsa@gmail.com. ORCID iD - 0000-0002-0811-1974

Correspondence:

Histopathologic & Immunohistochemical Analysis

Microscopic Features

The H&E stained sections revealed a biphasic pattern with areas of dense spindle cells denoted as Antoni A and loose, myxomatous stroma termed Antoni B. Characteristic palisading nuclei indicating Verocay Bodies were noticed. Large, pleomorphic, and hyperchromatic nuclei denoting Ancient change were scattered throughout the Antoni B zones which posed a challenge in diagnosis. These “smudged” nuclei appeared menacing but lacked mitotic activity. Dilated, thrombosed, and hyalinized blood vessels were prominent. [Figure 2a and 2b]

Special stains for mast cells with toluidine blue and immunohistochemistry were performed which shows uniformly distributed numerous mast cells. [Figure 2c] An intense and diffuse positivity for S100 Protein was noticed in the cytoplasm and nucleus of the spindle cells. A very low proliferation index with Ki-67 confirming the benign nature despite the atypia and negative SMA/Desmin ruled out smooth muscle tumors. [Figure 2d]

DISCUSSION

The term “Ancient Schwannoma” was coined by Ackerman and Taylor in 1951. The nuclear atypia seen in these cases is not a sign of malignant transformation but rather a degenerative process involving nuclear fusion and chromosomal derangement over time.

The diagnostic pitfall lies in the “pseudomalignant” appearance. Clinicians and pathologists must look for the “triple threat” of benignity that is presence of a capsule Verocay bodies and absence of atypical mitoses. [Table 1] Ancient schwannoma may show nuclear atypia mimicking malignancy such as Malignant Peripheral Nerve Sheath Tumor (MPSNST). But in this case, high mitotic activity, necrosis or infiltrative growth was not noticed. Mast cells are less prominent compared to benign lesions and reduced or patchy S-100 positivity

distinguishes ancient schwannoma from MPSNST.

Differential Diagnosis of Neurofibroma, Mastocytoma, Fibrous Histiocytoma was considered with relevance to degranulating mast cells. A focal distribution of mast cells in the present case rather than uniform distribution along with well-encapsulation and presence of Antoni A/B areas and Verocay bodies ruled out neurofibroma. Mast cells are commonly seen in schwannomas and are thought to promote tumor growth via cytokine release contribute to degenerative (“ancient”) changes. However, excessive degranulation can mimic mast cell-rich lesions like mastocytoma. Absence of features like sheets of mast cells rather than spindle cells, positive for tryptase, CD117 (c-KIT) and absence of Schwannian architecture excludes it. Fibrous Histiocytoma may contain inflammatory cells including mast cells, but it also does reveal a storiform pattern, Factor XIIIa positivity and lack of S-100 expression [3].

Spindle cell tumor with occasional mast cells like Leiomyoma can often pose as a diagnostic challenge, nevertheless intersecting fascicles of smooth muscle cells, SMA and desmin positivity and S-100 negativity eliminates the dilemma. Similarly, solitary fibrous tumor can show mast cells and spindle cells along with features like patternless architecture with “staghorn” vessels, STAT6 and CD34 positivity. Additionally, Traumatic Neuroma contains proliferating nerve fibers with mast cells along with distinguishing features such as disorganized nerve bundles, history of trauma or surgery and a mixed inflammatory infiltrate [4-8].

CONCLUSION

Neurilemoma of the lower lip is rare, and the Ancient variant is rarer still. Surgeons must maintain a high index of suspicion when clinical findings (slow growth, encapsulation) do not match alarming histopathologic reports of atypia. Conservative surgical excision remains the gold standard, with an excellent prognosis and minimal recurrence.

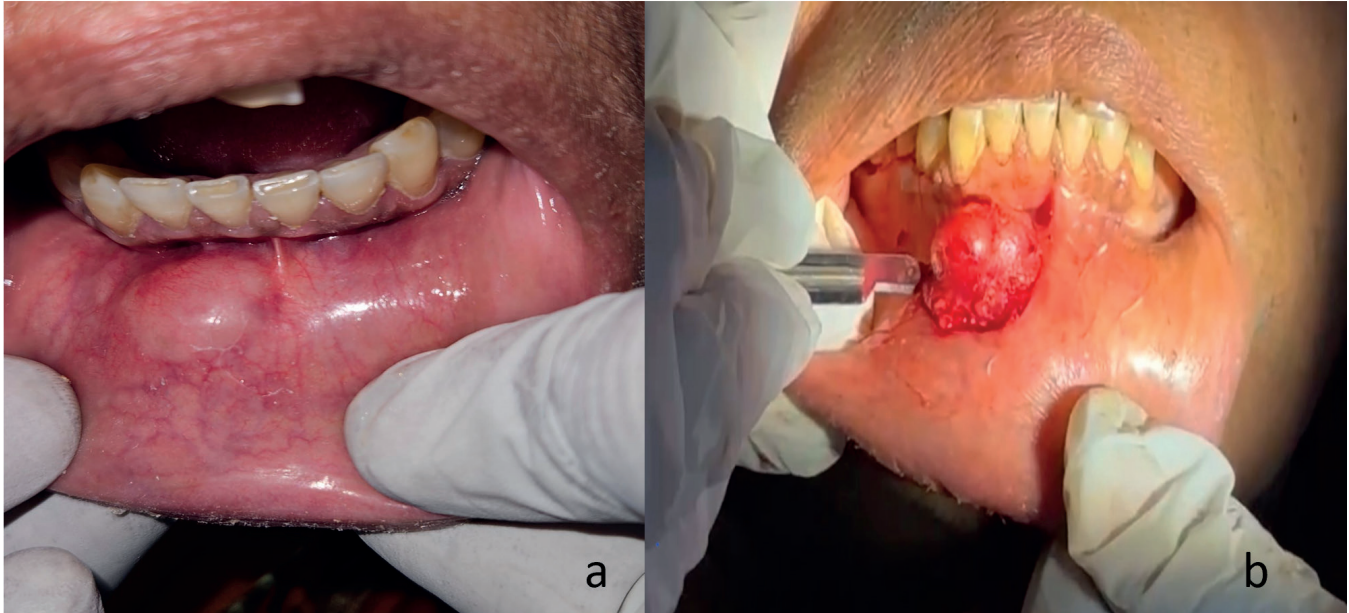


Figure 1: Clinical photograph showing the submucosal nodule on the lower lip.

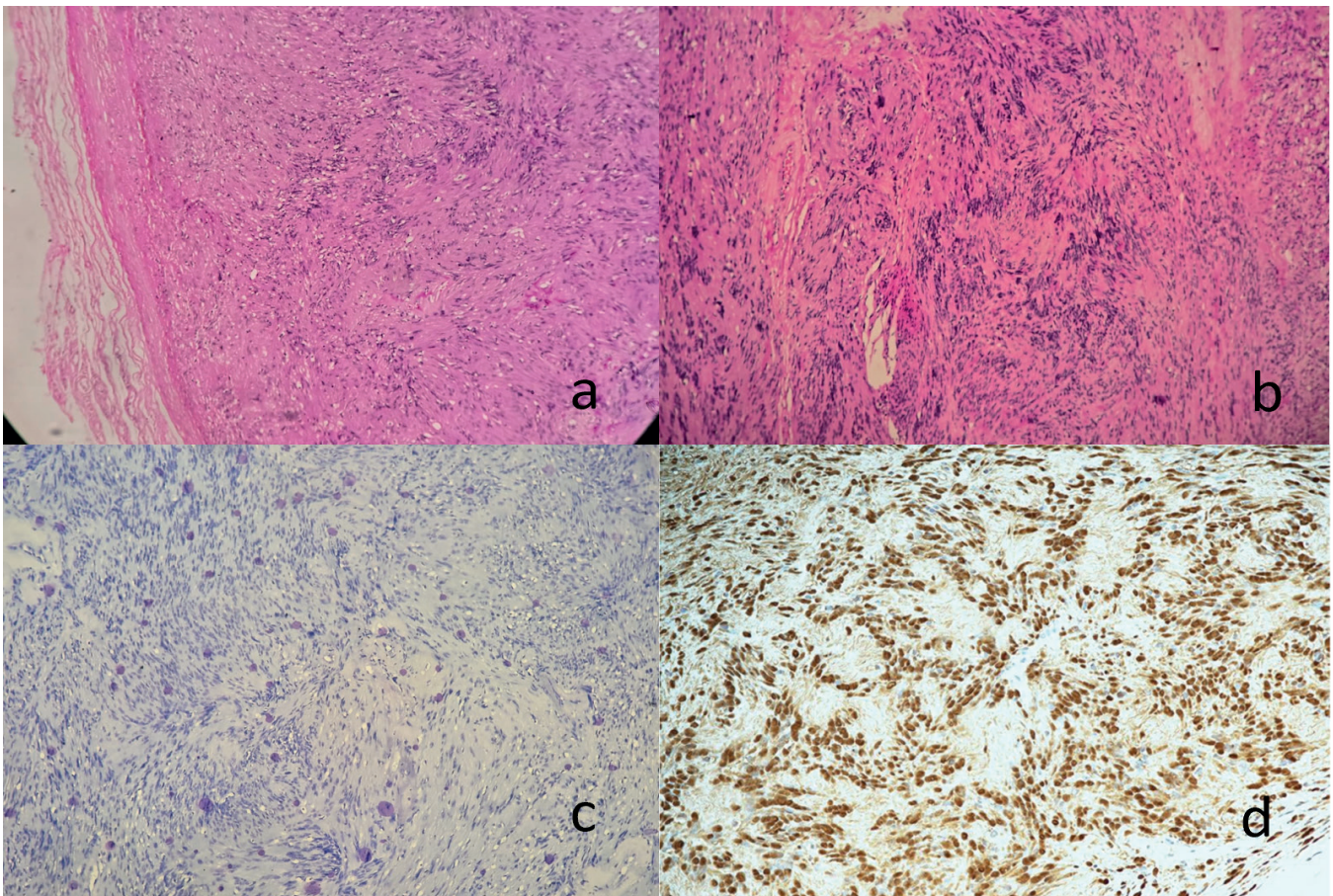


Figure 2: (a) H&E (10x) showing the capsule and the transition between Antoni A and Antoni B areas. (b) H&E (40x) highlighting the Ancient changes: large, irregular, dark nuclei within a hypocellular stroma. (c) Toluidine blue stain showing numerous mast cells. (d) IHC showing strong, diffuse S100 expression.

Table1: Differential diagnosis of Spindle cell lesions with mast cells

Feature	Ancient Schwannoma	Neurofibroma	MPNST	Mastocytoma
Encapsulation	Present	Absent	Absent	Absent
Mast cells	Abundant, degranulating	Numerous, uniform	Few	Predominant
Cell type	Schwann cells	Mixed cells	Atypical spindle	Mast cells
Antoni A/B	Present	Absent	Absent	Absent
Verocay bodies	Present	Absent	Absent	Absent
Atypia	Degenerative only	Minimal	Marked	Absent
Mitosis	Absent	Rare	Frequent	Absent
Necrosis	Absent	Absent	Present	Absent
S-100	Strong diffuse +	Variable	Reduced/patchy	Negative
Key point	Atypia without mitosis	Mixed cellularity	Malignant features	Sheets of mast cells

REFERENCES

- Ackerman LV, Taylor FH. Neurogenous tumors within the thorax. A clinicopathological evaluation of forty-eight cases. *Cancer*. 1951;4(4):669-691.
- Sreeja C, et al. Ancient schwannoma of the cheek: A case report and review. *Journal of Oral and Maxillofacial Pathology*. 2015;19(2):270.
- Scurry WC, et al. Ancient schwannoma of the parapharyngeal space. *Archives of Otolaryngology–Head & Neck Surgery*. 2006;132(11):1258.
- Priyanka S, et al. Neurilemmoma of the lower lip: A rare entity. *Case Reports in Dentistry*. 2015.
- Vong KS, Mohamad I, Jaafar R. Neglected left intraparotid facial nerve schwannoma causing complete facial nerve palsy: a case report. *Bangladesh J Med Sci*. 2018;17(4):680-682. doi:10.3329/bjms.v17i4.38338.
- Irfan M, Halim SA, Khir AM. Infratemporal neurofibroma presenting as a parotid mass. *Bangladesh J Med Sci*. 2009;8(3):72-74. doi:10.3329/bjms.v8i3.3987.
- Alam MK, Alhabib S, Alzarea BK, Irshad M, Faruqi S, Sghaireen MG, et al. 3D CBCT morphometric assessment of mental foramen in Arabic population and global comparison: imperative for invasive and non-invasive procedures in mandible. *Acta Odontol Scand*. 2018;76(2):98-104. doi:10.1080/00016357.2017.1387813.
- Aloyouny A, Albagieh HN, Alweteid AS, Alsheddi MA, Alam MK. Mandibular neurofibroma with osseous deformities in a 3-year-old child with neurofibromatosis type 1: a case report presentation and diagnosis. *Niger J Clin Pract*. 2021;24(11):1709-1713.