

Case Report



A Case Report of a Huge Nerve Sheath Myxoma of the Anterior Gingiva in a 47-Year-Old Lady: Diagnostic Confirmation with Immunohistochemical Analysis

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Abstract

Background: Nerve sheath myxoma (NSM) is a rare, benign soft tissue neoplasm arising from peripheral nerves. It typically presents as a slow-growing, painless mass, often in the extremities. Intraoral NSMs are exceedingly rare, and large lesions involving the gingiva are even less common.

Case Presentation: This case report describes a 47-year-old female who presented with a large, exophytic, painless mass involving the anterior gingiva. The lesion was surgically excised, and histopathological examination, together with immunohistochemical staining for S-100, EMA, Calponin, and SOX-10, confirmed the diagnosis of nerve sheath myxoma.

Conclusion: This report highlights the rarity of NSM, its diagnostic challenges, and the importance of immunohistochemistry for accurate diagnosis, particularly at unusual intraoral sites.

Keywords: Nerve sheath myxoma, S-100 protein, Neurothekeoma, Calponins, Immunohistochemistry

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Introduction

Myxoid tumors of the oral cavity encompass a broad spectrum of lesions, primarily neoplastic.^{1,2} Nerve sheath myxoma (NSM), also known as neurothekeoma of the nerve sheath, is a benign, uncommon soft tissue neoplasm originating from peripheral nerve sheaths. It was first described by Gallager and Soule in 1976.³ NSM commonly presents as a solitary, slow-growing, painless mass, typically in the extremities, particularly the upper extremities and shoulders. Less frequently, however, NSMs can arise in the head and neck region, including the oral cavity.^{3,4} Intraoral NSMs are extremely rare, accounting for a small proportion of all reported cases.⁵ When present in the oral cavity, they tend to involve the tongue, buccal mucosa, and palate.⁶ Gingival involvement, particularly by large lesions, is exceedingly rare, making this case of significant interest.

The diagnosis of NSM can be challenging because of its variable histological presentation and similarities to other soft tissue tumors. Histopathologically, NSM is characterized by a myxoid stroma containing spindle-shaped or stellate cells, often accompanied by small blood

vessels.⁶ Immunohistochemical staining plays a crucial role in confirming the diagnosis. S-100 protein, a marker of neural crest-derived cells, is frequently positive in NSMs.⁷ Epithelial membrane antigen (EMA) positivity may also be observed, whereas markers such as calponin and SOX-10 are important for differentiating NSM from other morphologically similar lesions.

This case report presents a rare instance of a large NSM located on the anterior gingiva of a 47-year-old female, emphasizing the diagnostic importance of histopathological examination and immunohistochemical analysis, particularly the use of S-100, EMA, Calponin, and SOX-10 for an accurate diagnosis.

Case Report

A 47-year-old female presented to the clinic with a painless, slowly growing mass on the anterior gingiva. The patient reported that the lesion had been present for approximately 1 year and had gradually increased in size. Her medical history was non-contributory, and she denied any previous history of trauma or surgery in the oral cavity.



On clinical examination, a large, exophytic, soft, non-ulcerated mass was observed extending from the attached gingiva between the maxillary right central incisor and canine (Figure 1).

The lesion measured approximately 2.5 x 2.0 x 1.5 cm. The overlying mucosa was smooth and appeared normal in color. Palpation revealed a soft, slightly compressible mass without tenderness or pain. There was no evidence of regional lymphadenopathy. The patient reported no functional limitations secondary to the lesion. T2 weighted MRI revealed a large hyperintense area in the anterior mandible measuring about 6.09 cm x 1.43 cm (Figure 2)

Based on the clinical presentation, the differential diagnosis included fibroma, pyogenic granuloma, peripheral giant cell granuloma, and peripheral ossifying

fibroma.

An excisional biopsy was performed under local anesthesia. The lesion was carefully removed along with a small margin of surrounding normal tissue to ensure complete excision. The surgical defect was closed primarily with sutures (Figure 3)

Histological examination of the excised specimen revealed a well-circumscribed, unencapsulated lesion within the connective tissue of the gingiva. The lesion was characterized by a predominantly myxoid stroma containing loosely arranged, bland-appearing spindle-shaped and stellate cells with wavy nuclei (Figure 4).

Areas of increased cellularity and collagen deposition were noted. Small, thin-walled blood vessels were scattered throughout the stroma. No evidence of mitotic activity,

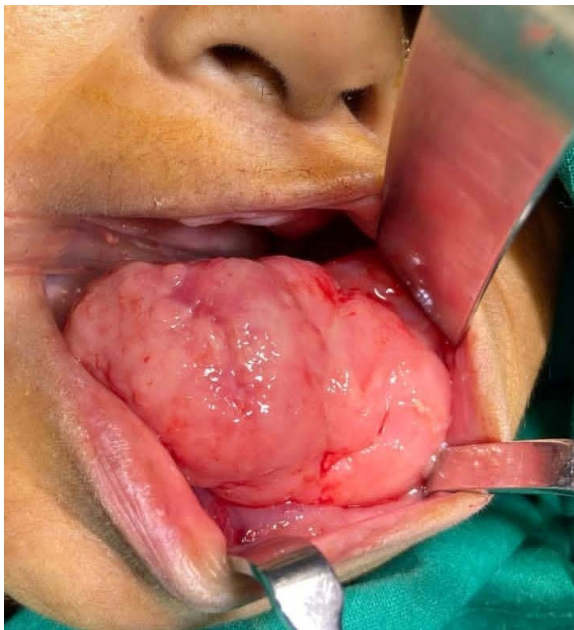


Figure 1. Clinical picture of the lesion



Figure 2. MRI of the lesion

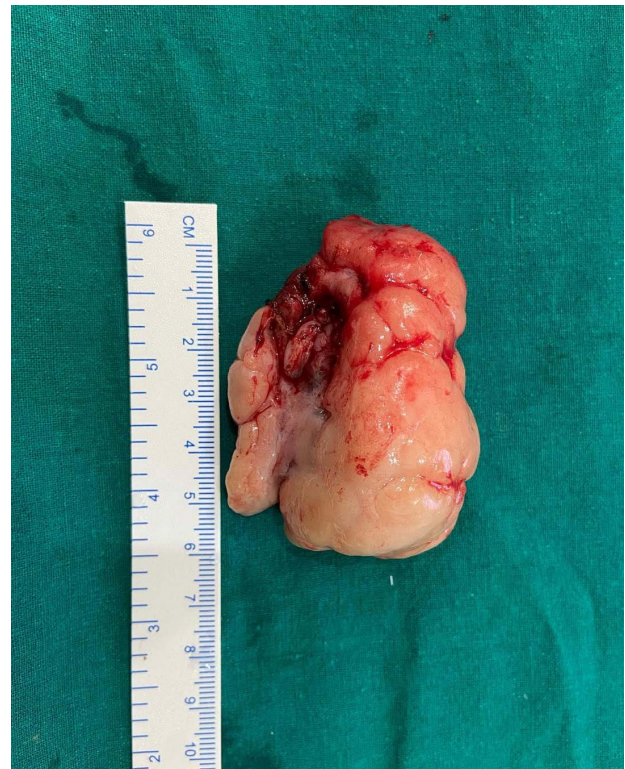


Figure 3. Gross picture of the lesion after removal

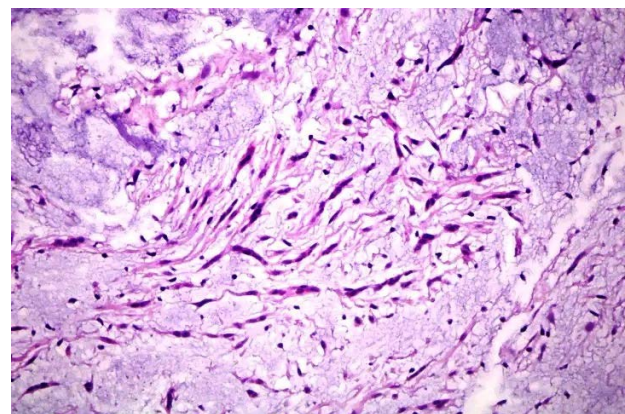


Figure 4. Collection of numerous spindle- and stellate-shaped cells in a predominantly myxoid stroma (Hematoxylin and Eosin Original magnification X40)

necrosis, or atypia was observed. To confirm the diagnosis and differentiate NSM from other soft tissue neoplasms, immunohistochemical analysis was performed. The analysis revealed strong positivity of the spindle cells for S -100 (Figure 5a); EMA was positive in a subset of cells (Figure 5b); Calponin was negative (Figure 5c); and SOX -10 showed nuclear positivity in the spindle shaped cells (Figure 5d)

Based on the histopathological and immunohistochemical findings, a diagnosis of nerve sheath myxoma was confirmed.

The patient was followed up for 6 months, during which no recurrence of the lesion was observed. The surgical site healed well, and the patient reported no complications.

Discussion

Nerve sheath myxoma is a rare, benign neoplasm of peripheral nerve origin. Its occurrence in the oral cavity, particularly on the gingiva, is exceptionally rare (Table 1)

In the oral cavity, the first case of NSM was reported by Prado JD et al in 2007. The clinical presentation of NSM is typically a slow-growing, painless mass that can mimic

more common benign lesions of the oral cavity. In this case, the initial differential diagnosis included common lesions such as fibroma, pyogenic granuloma, peripheral giant cell granuloma, and peripheral ossifying fibroma.⁶

The definitive diagnosis of NSM relies on histopathological examination and immunohistochemical analysis. Histologically, NSM is characterized by a myxoid stroma containing spindle-shaped and stellate cells. The presence of small blood vessels is also a characteristic feature. The immunohistochemical profile of the lesion is

Table 1. Description of NSM cases reported in the gingiva

S.NO	Age	Gender	Size of the lesion	Immunohistochemical interpretation.	Authors
1	84	F	1 cm	Strong positivity for Vimentin and S -100	Prado JD et al.(5)
2	32	F	0.8×0.6×0.6 cm	Sporadically positive for S -100. Negative for GFA, EMA, SMA, CD 68 and HMB 45	Safadi RA et al.(9)
3	11	M	2×1.5 cm	NA	Bajpai M et al.(8)

(F –Female, M – Male, NA –Not available)

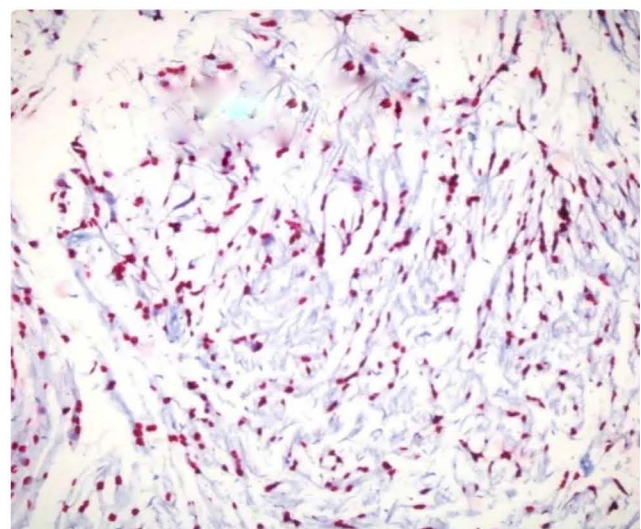
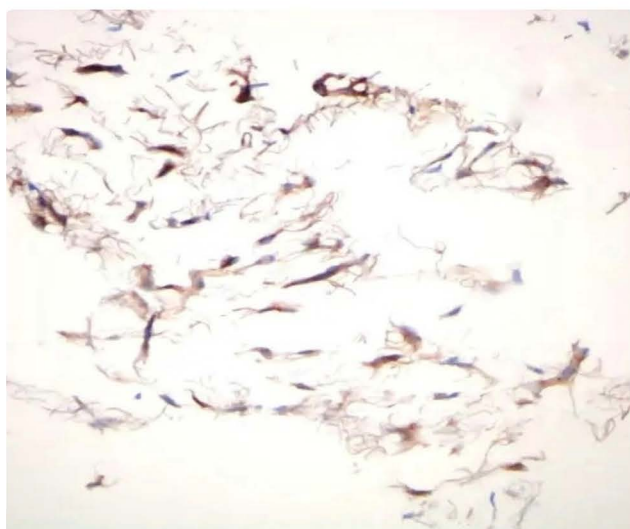
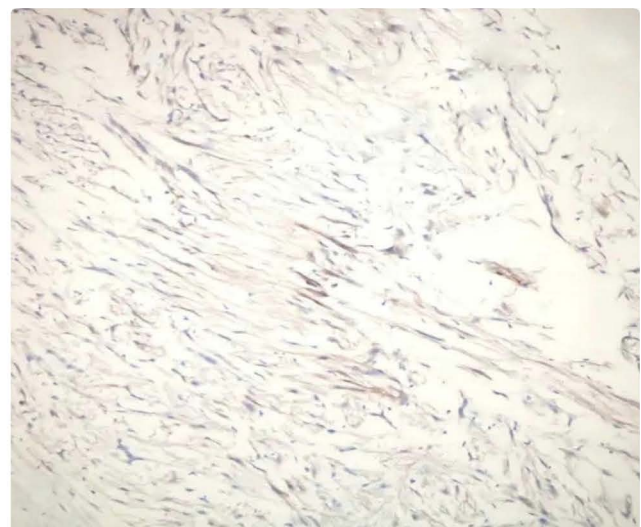
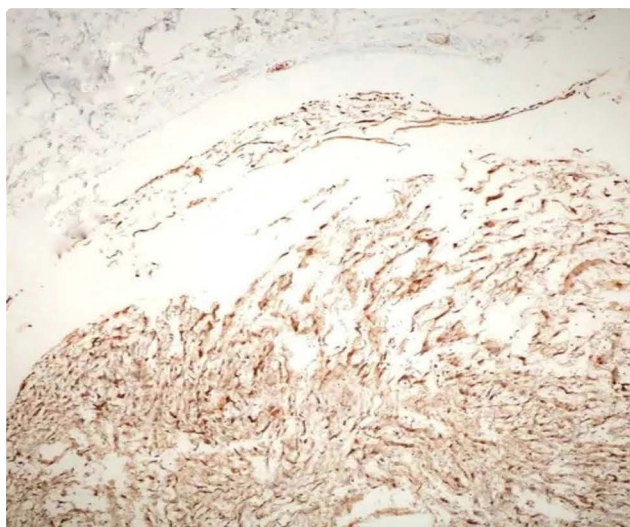


Figure 5. Positive expression of S -100 in the spindle cells (a) Negative expression of calponin in the lesional cells (b) Focal positivity of EMA in the cells (c) Positive expression of SOX -10 in the nuclei of spindle shaped cells (d)

key to confirming the diagnosis.⁷

S-100 protein is a widely used marker of neural crest-derived cells and is frequently positive in NSMs.⁸ However, S-100 positivity is not specific to NSM and can also be observed in other tumors, such as schwannoma, neurofibroma, and melanoma. EMA positivity, often showing a perineurial pattern, may also be observed in NSMs.⁹ Calponin, a marker of smooth muscle differentiation, is usually negative in NSMs, which helps differentiate NSM from lesions with smooth muscle components. Recently, SOX-10 has emerged as a highly sensitive and specific marker for nerve sheath tumors, including NSM.^{9,10} SOX-10 is a transcription factor involved in neural crest development and differentiation. Its positivity in the nuclei of spindle cells further supports the diagnosis of NSM.

The differential diagnosis of NSM includes other myxoid lesions, such as myxoma, schwannoma with myxoid degeneration, and low-grade fibromyxoid sarcoma. Immunohistochemical analysis is crucial for differentiating these lesions. Myxomas are typically negative for S-100 and SOX-10. Schwannomas are strongly positive for S-100 but may exhibit histopathological features that differ from those of NSM. Low-grade fibromyxoid sarcoma is characterized by a more infiltrative growth pattern and may show positivity for MUC4.¹¹⁻¹⁶

The primary treatment for NSM is surgical excision with a small margin of normal tissue. Given the benign nature of the tumor, conservative excision is usually sufficient. Although recurrences are rare, long-term follow-up is recommended to monitor for potential recurrence.¹¹

This case highlights the importance of considering rare entities such as NSM in the differential diagnosis of oral lesions, particularly those presenting as slow-growing, painless masses. Accurate diagnosis requires careful histopathological examination and the judicious use of immunohistochemical markers, including S-100, EMA, Calponin, and SOX-10.

Written consent was obtained from the patient for publication of this case.

Conclusion

This case describes a rare presentation of a large nerve sheath myxoma located on the anterior gingiva. The diagnosis was confirmed by histopathological examination and immunohistochemical analysis, demonstrating the importance of these diagnostic tools in differentiating NSM from other morphologically similar lesions. Positive staining for S-100, EMA, and SOX-10, together with negative staining for Calponin, provided strong evidence supporting the diagnosis of NSM. Surgical excision with clear margins was effective in treating the lesion, with no recurrence observed during the follow-up period. This case emphasizes the need for clinicians to consider rare entities such as NSM in the oral cavity and to use appropriate diagnostic techniques for accurate diagnosis and management.

Artificial Intelligence Use Disclosure

Authors declare that they have not used AI in preparing this manuscript.

Authors' Contribution

Conceptualization: Deepak Singh, Vandana Pandey Tripathi
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Investigation: Manas Bajpai, Saurabh L Sabnis
Methodology: Deepak Singh
Project Administration: Vandana Pandey Tripathi
Software: Saurabh L Sabnis
Supervision: Saurabh L Sabnis, Manas Bajpai
Resource: Saurabh L Sabnis
Validation: Deepak Singh
Visualization: Deepak Singh
Writing–Original Draft: Deepak Singh
Writing–Review & Editing: Saurabh L Sabnis

Competing Interests

We have no competing interests.

Data Availability Statement

As the present manuscript is a case report, a data availability statement is not applicable.

Ethical Approval

This manuscript was ethically approved by the Institutional Ethical Committee.

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