MUCOCELE ON LOWER LIP? NOT ALWAYS! A CASE REPORT

Dr Seema M.1, Dr Leeky Mohanty2, Dr Shruti Srinivasan3, Dr Saleha Jamadar4, Dr Komali Y5

1Senior Lecturer, Department of Oral and Maxillofacial Pathology
2HOD, Department of Oral and Maxillofacial Pathology
3Senior Lecturer, Department of Oral and Maxillofacial Pathology
4Reader, Department of Oral and Maxillofacial Pathology
5Reader, Department of Oral and Maxillofacial Pathology

Conflicts of Interest: Nil
Corresponding author: Dr Seema M
DOI: https://doi.org/10.32553/ijmsdr.v5i2.758

Introduction:
Lesions of the lip that are smooth, spherical, and with same color of the oral mucosa are often diagnosed as traumatic fibromas, or focal fibrous hyperplasia, and mucoceles.1 Although tumors of the peripheral nerve sheath are uncommon, they must also be included in the list of differential diagnoses of lesions that are nodular involving lower lip. Peripheral Nerve Sheath Tumors (PNST) are rare lesions of the oral cavity. They comprise of reactive to neoplastic lesions that arise due to proliferation of axons, Schwann cells or their lining structures. PNSTs are classified based on their cellular composition and organization. Schwannoma, Neurofibromas, Palisaded Encapsulated Neuroma, Traumatic Neuroma, Nerve Sheath Myxomas, Perineurioma, Granular Cell Tumor, are few of the PNSTs reported apart from malignant nerve sheath tumors.2 Palisaded Encapsulated Neuroma (PEN) is a distinctive neural tumor of benign origin, often presents as small, solitary, firm, asymptomatic, rubbery, skin-colored nodules or papule. It is most commonly seen in middle-aged adults with male to female ratio of 1:1. It rarely represents with multiple lesions.3

Case Report
A 45 year old male patient presented to the hospital for routine dental examination. On examination a soft swelling was noted on left vermilion border of lower lip extending on to labial mucosa measuring about 0.5 * 0.5 cm which was asymptomatic and appeared normal in color similar to adjacent mucosa. No history of trauma was reported. No relevant medical history reported. A provisional diagnosis of mucocele was given. An excisional biopsy was performed. On histopathological examination, a well circumscribed and partially encapsulated connective tissue was seen. Centrally placed axons surrounded by short interlacing fascicles of palisading spindle shaped cells with elongated wavy nuclei, suggestive of Schwann cells were seen. There was no evidence of mast cells, mitosis and pleomorphism. A lining of parakeratinated stratified Squamous epithelium was present. Based on the above findings a histopathological diagnosis of Palisading Encapsulated Neuroma was rendered.

Figure 1: Soft swelling noted on the left vermilion border of lower lip
Figure 2: Lesion noted extending on to labial mucosa with color of the lesion similar to that of adjacent mucosa
**Discussion:**

In 1972, PEN of the skin was first described by Reed et al.\(^4\) It was described as a distinctive lesion which is usually small, solitary and asymptomatic often skin colored resembling a papule that arises mostly on the face or on the mucocutaneous junctions. It is a rare slow growing benign neural tumor, with variable size of 2 to 6 mm often detected during the 5th to 7th decade of life.\(^6\) frequently misdiagnosed as neurofibroma, melanocytic nevus, basal cell carcinoma, epidermal cyst, or skin appendage tumor.\(^5\)

Intraoral PEN have dome shaped growth with smooth surface with hard palate being the most commonest site. The overlying mucosa has similar color to the adjacent region.

The etiopathogenesis of PEN had been a subject of debate. It was proposed that the lesion is an attenuated manifestation of multiple endocrine neoplasia type 2B (MEN2B). MEN2B is characterized by development of neoplasms in the endocrine glands eyes and mouth. Nevertheless, distinct differences existed in the histopathological features of PEN to that of lesions associated with MEN2B. Trauma had also been proposed as an etiological factor. However, most cases of PEN does not exhibit a history of trauma.\(^7\)

Conversely, the tongue being a common site of oral trauma, is infrequently affected. Tongue, lips as well as the palate and gingival contain nerve branches that are superficially situated compared to the buccal mucosa. Infrequently, PEN do include areas that can be encountered in traumatic neuroma.\(^8\)

Histopathologically, it has a appearance between that of a schwannoma and a neurofibroma. It is essential to differentiate Neurofibromas from PEN, as neurofibromas are associated with neurofibromatosis, and has a predilection for malignant transformation.\(^9\)

PEN consists of partially encapsulated mass of moderately cellular interlacing fascicles of spindle cells that are consistent with Schwann cells. Nodular, epithelioid, plexiform and multinodular type of

**Figure 3:** A well circumscribed and partially encapsulated connective tissue seen along with a lining of stratified Squamous epithelium.

**Figure 4:** Short interlacing fascicles of palisading spindle shaped cells with elongated wavy nuclei, suggestive of Schwann cells seen. There was no evidence of mast cells, mitosis and pleomorphism.

**Figure 5:** There was no evidence of mast cells, mitosis and pleomorphism.

**Figure 6:** Centrally placed axons surrounded by short interlacing fascicles noted.
presentation can also be observed with nodular pattern being the commonest type. Tumor cells are poorly delineated with eosinophilic cytoplasm with basophilic nuclei that are characteristically wavy and pointed with no significant pleomorphism or mitotic activity similar to our case. Based on the clinical presentation and histopathologic features, traumatic neuroma is the closest differential diagnosis that can be considered for the present case. Traumatic neuroma is a hyperplastic response of the nerve tissue with history of trauma in the region. The lesion is characterized by firm nodular growth with severe tenderness in the region and microscopic evidence of perineural cells rimming discrete microfascicles and inflammatory cells which was not present in our case. Schwanoma being another differential diagnosis was ruled out microscopically as Verocay bodies and nuclear palisading were absent. Cunha et al reported a case of intraoral PEN mimicking mucocele of lower labial mucosa similar to our case however microscopically had no evidence of salivary gland pathology. Neurofibroma is non encapsulated and shows hypocellular sheets, mucoid matrix with delicate collagen and significant number of mast cells which were absent in our case. Some pathologists prefer solitary circumscribed neuroma(SCN) as a better descriptive term for lesion that is always not encapsulated and cells are usually not truly palisaded like in PEN which was not seen in our case. The tumor cells of PEN show reactivity to S100 protein. However unlike other neural tumors they are negative for GFAP. Prior to general Oral And maxillofacial pathologists becoming familiar with PEN/SCN, lesions were variously reported as neurofibromas, schwannomas, or lesions that were intermediate between the two. A review on oral spindle cell tumors, revised 15 original diagnoses of schwannoma and neurofibroma to PEN/SCN in 2003. However, we rarely encounter challenges in distinguishing PEN/SCN from other benign peripheral nerve tumors. Incidence of mucocele is 0.4-0.9% in general population and it is the 17th most common lesion in the oral cavity with lower lip being most commonly affected (36.20%). Mucoceles are asymptomatic in most cases, with similar other clinical features of PEN becomes physicians’ main choice as a provisional diagnosis. PEN comprise 4.5% of oral soft tissue neoplasms. Although tumors of the peripheral nerve sheath are uncommon, they must also be included in the list of differential diagnoses of lesions that are nodular involving lower lip. Familiarity with clinical, histopathological and immunohistochemical aspects of such lesions are essential to avoid misdiagnosis and render a satisfactory therapeutic approach to patients. Complete surgical excision is the treatment of choice for PEN. Similarly complete surgical excision was performed in our case and follow up for 2 months showed no recurrence of the lesion.

Table 1: Average of Demographic Data including Gender and age

<table>
<thead>
<tr>
<th>Gender</th>
<th>Male: Female ratio (%)</th>
<th>60:40%</th>
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<tbody>
<tr>
<td>Age</td>
<td>Average Age</td>
<td>46yrs</td>
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Table 2: Average of Distribution of Site of the lesion

<table>
<thead>
<tr>
<th>Site of the lesion</th>
<th>Percentage</th>
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<tbody>
<tr>
<td>Palate</td>
<td>61.2%</td>
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<tr>
<td>Upper Lip</td>
<td>15.5%</td>
</tr>
<tr>
<td>Tongue</td>
<td>12.2%</td>
</tr>
<tr>
<td>Lower Lip</td>
<td>8.5%</td>
</tr>
<tr>
<td>Gingiva</td>
<td>11.4%</td>
</tr>
<tr>
<td>Buccal Mucosa</td>
<td>4.1%</td>
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Table 3: Average of the Duration of the lesion\textsuperscript{15}

<table>
<thead>
<tr>
<th>Duration</th>
<th>0-5yrs</th>
<th>15.7%</th>
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<tr>
<td>6-10yrs</td>
<td>15.7%</td>
<td></td>
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<tr>
<td>&gt;11yrs</td>
<td>15.7%</td>
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Table 4: Average of the clinical diagnosis with Histopathological confirmed cases of PEN\textsuperscript{15}

<table>
<thead>
<tr>
<th>Clinical Diagnosis</th>
<th>Papilloma</th>
<th>15.7%</th>
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<td>Fibroma</td>
<td>83.3%</td>
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References