A Nerve Sheath Myxoma (Soft Tissue Myxoma) of the Lower Lip: Rare Case Study

Rana F. Swaidan*

ABSTRACT

Myxomas in the oral cavity and lips are relatively uncommon tumors that present in different types: the first type is odontogenic myxomas and the second type is soft tissue myxomas. Odontogenic myxomas (originated from the mesenchymal cells of the oral cavity such as dental papillae, periodontium, or dental follicles) are most presented in the maxilla and mandible. In comparison, soft tissue myxomas are rare in the oral cavity and lips, and only 30 cases of unknown development were reported in the literature, and only five cases involving the lip were reported. Even if it is considered very rare, nerve sheath (soft tissue) myxoma must be included in the differential diagnosis of lip lesions and excised widely to prevent local recurrence. The aim of our case study is to present a rare case of lip myxoma in a male patient 26-year-old who presented with a soft, exophytic lesion on the lower lip.

Keywords: Lip, myxoma, nerve sheath, soft tissue.

1. Introduction

Nerve sheath myxoma (neurothekeoma), is identified as a benign tumor of nerve sheath. Which was described initially as a cutaneous lesion with benign behavior and a pattern of neural growth which gave it initially the name nerve sheath myxoma followed later by the name Neurothekeoma [1]. Nerve sheath myxoma mostly seen on the head and neck region but rarely in the lower lip [2], [3].

2. Case Report

A young male 26-year-old patient was referred to our department of maxillofacial surgery at Al-Basheer Hospital from a general dental clinic in January 2023. He was complaining of a painless asymptomatic mass that had been in the lower lip for one year previously.

Clinical examination showed a 1.5 cm soft, round exophytic lesion (mass) located in the lower lip (Fig. 1). The lesion was painless not tender covered by normal oral mucosa extended beyond the vermillion border to the skin of the lower lip. The patient has no medical problems, and his dental history has no abnormalities. Under local anesthesia, surgical removal was carried out by a plastic surgeon 1 year ago with the diagnosis of a rupture inclusion cyst. After 3 months the lesion recurred and the patient was referred to our clinic for consultation, a biopsy (incisional type) was taken under local anesthesia. The incised specimen was sent for histopathological evaluation. The report came with a diagnosis of nerve sheath myxoma (soft tissue myxoma).

Microscopically, the lesion was composed of epithelioid cells along with spindle-shaped cells that were loosely arranged with strange myxoid stroma seen after Hematoxylin and Eosin staining (Fig. 2). For final diagnosis.

Fig. 1. Clinical examination showed the soft and round lesion 1.5 cm in diameter on the lower lip.
A Nerve Sheath Myxoma (Soft Tissue Myxoma) of the Lower Lip: Rare Case Study

Swaidan

Immunohistochemistry stains were done with positive CD 63 and CD 34 in blood capillaries, and negative for Desmin and S-100 protein.

The final diagnosis of soft tissue myxoma was made based on the histopathological findings. A decision was taken for wide excision to prevent recurrence making a V-shaped resection of the lesion followed with primary closure.

Six months and Nine months follow-up after the operation, no recurrence of the mass was detected (Fig. 3).

3. Discussion

In the head and neck regions, odontogenic type and soft tissue type are considered the forms of myxoma seen. Odontogenic types are benign growths originating from mesenchymal cells such as dental papillae, or periodontium, which present more frequently than soft tissue counterparts in both jaws (maxilla and mandible) [1]. Peripheral (soft tissue) myxomas are a group of contrastive soft tissue tumors with a similar histological presentation of copious myxoid material in their background [1], [4]. Soft tissue myxomas are considered a growth of primitive undifferentiated mesenchymal cells of perineural origin or nerve cell origin [2], [3]. There are a wide range of groups of soft tissue tumors with different invasion properties, ranging from benign to highly aggressive types [5]. Although they are mostly benign, the locally infiltrative nature makes recurrence a common feature if the excised margins are positive [4], [5].

Soft tissue myxomas are presented and reported in different parts of the head and neck regions for example in skeletal muscles, subcutaneous tissues, and different parts of the oral cavity [6]. several theories of its pathogenesis were suggested but its origin remains unclear till the present day [7].

The clinical features of nerve sheath myxomas seen in the oral cavity are not pathognostic as reported in the literature. females are more commonly affected than males [8]. it affects a wide range of ages from infants a few weeks old until elderly in their 8th decade of life, however, most of the cases are seen in the 4th decade. The frequent symptom reported in most patients was a painless mass or nodule slowly growing over a long period of time ranging from months to several decades [2], [7]. The differential diagnosis for soft tissue myxoma is lipoma, fibroepithelial polyp, fibroma, and salivary gland tumor [8], [9]. Histopathological examination with immune stains is the only way to get a precise diagnosis of these lesions [1], [2].

The most frequent site of involvement in the oral cavity was the palate, the next site was the buccal mucosa, followed by the lips and the floor of the mouth according to a report done by [1] and [10]. Nisi et al. [9] reported more cases of nerve sheath myxomas given the palate followed by buccal mucosa the most frequent sites. Mason et al. reported the first soft tissue myxoma involving the lip [2], [11]. Only five cases including the lips were reported in the literature [2].

Histological examination of nerve sheath myxomas using Hematoxylin and Eosin stains shows a lesion with hypocellularity which consists of epithelioid cells along with spindle-shaped cells immersed in excess myxoid or mucous background that has fibers (mainly reticulin type) [4], [11]. In General, nerve sheath myxoma is categorized into three different types according to the myxoid background of the lesion, they are classical type, cellular type, and mixed type [2]. Special staining techniques and immunohistochemistry stains like CD34, SMA, CD63, S100, and desmin are of great help in diagnosing and identifying lesions from similar entities [1], [5]. In this case study, the definitive diagnosis of nerve sheath myxoma was made after a complete histological examination and immunohistochemistry staining results since the clinical presentation of such a lesion is not pathognomonic.

Surgery is considered the first option for treating nerve sheath myxomas where wide local excision with adequate safety margins (about 1 cm) is performed to prevent local recurrence since recurrence is greatly associated with inadequate margin excision. However, if vital structures will be involved in wide surgical resection a more conservative
approach is used to preserve them. Recurrence of nerve sheath myxoma ranges between 5%–30% which greatly depends on the type of treatment and surgical margins, but luckily none of the oral cavity soft tissue myxoma reported had any recurrence but close follow-up is advised [2], [7].

4. Conclusion

Our case study describes both clinical and histological findings of soft tissue myxoma (nerve sheath type) of the lower lip which mainly presented as a painless benign soft tissue lesion (mass) on the lower lip and depending on its clinical behavior and potential recurrence a wide local excision with adequate safety margins is recommended as primary treatment. Although nerve sheath myxoma of the lower lip is considered a rare entity, it should be always considered in the differential diagnosis of lower lip masses.

Conflict of Interest

Author declares no conflict of interest.

REFERENCES