

A Rare Case Of A Large Intraoral Neurothekeoma Involving Mental Nerve: A Case Report.

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Abstract:

Neurothekeoma, a lesion of the nerve sheath, have been reported to be a rare benign lesion which is usually asymptomatic and slow growing. These lesions are difficult to diagnose prior to any biopsy and histopathological analysis. The treatment being complete excision of the lesion, with rare chances of recurrence, which otherwise might be due to incomplete excision. Here a case has been presented with Neurothekeoma involving the mental nerve with a follow up of 6 months, post excision. The healing has been uneventful with no signs of recurrence. This case report might be a step towards creating more awareness about the lesion along with its diagnostic differentiation from other lesions and the treatment aspect.

Keywords: Neurothekeoma, nerve sheath, benign lesion

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I. Introduction:

Neurothekeomas are described as a rare benign lesions of nerve sheath that is typically present as asymptomatic nodules on the face and neck in children and young adults with a slight female predominance. It was initially described by Hakin and Reed, in 1969, as Nerve sheath myxoma.¹ This tumor usually involves dermis or, less frequently, mucosal or submucosal tissue and, typically, is described as a small, solitary, slow growing and reddish to flesh-colored nodule or papule. The lesions usually are found as solitary skin-colored nodules, 90% measured <2 cm.² Histologic variants include myxoid, cellular and mixed tumors.

Usually, neurothekeomas are difficult to diagnose preceding a biopsy, which may be due to lack of specific clinical manifestations or imaging characteristics.⁹

II. Case Report:

Here we present a case (Male/51 years) that had a chief complaint of swelling in his chin and lower left facial area. The swelling, as the patient reported and as per he had noticed, was present since 8-9 years and had gradually increased from a small size to the present size. No pain or discomfort was reported, and only esthetic dissatisfaction was expressed by the patient. Examination revealed skin colored solitary mass with firm consistency and slight tenderness on palpation was reported by the patient. A decision for incisional biopsy was proposed for final diagnosis and treatment planning.

Histological examination revealed stratified squamous epithelium and an underlying lamina propria with proliferating tumor mass was arranged as lobules. The slide showed ovoid/spindle cells and epithelioid cells with bland, vesicular nuclei, and a light eosinophilic cytoplasm. The diagnosis of neurothekeoma was given. The patient was advised for IHC (immunohistochemical analysis) but due to unexplainable circumstances the patient was unable to do the same. Thus it was decided for a complete surgical excision of the lesion.

The procedure was carried out in proper supervision. An intraoral incision from left lower canine region to 2nd molar region was given. Submucoal dissection was carried out to expose the lesion. After the dissection, it was observed that there was presence of well capsulated mass attached to the mental nerve. The complete excision of the lesion was carried along with sacrificing the mental nerve.

The lesion roughly measured around 9cm x 4.5cm which to the best of our knowledge is a neurothekeoma lesion of pretty large size in the oral cavity, ever to be published in other literature or case reports.

A 6 month follow up of the patient showed no uneventful recurrence at the site of surgery and no other complaints from the patient.

List Of Figures:



Fig 1: Facial profile of patient with a well defined swelling extending from lower left symphysis region to body of mandible

Fig 2: Intraoral incision given, revealing a well encapsulated lesion involving the mental nerve

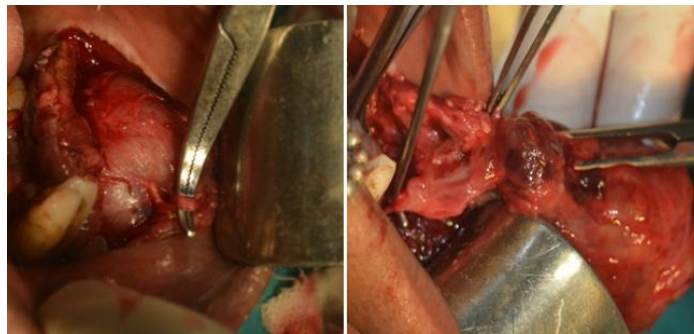


Fig 3: The mental nerve within the lesion

Fig 4: The lesion being excised



Fig 5: the excised lesion measuring around 9 x 4.5cm

Fig 6: Intraoral site after excision of the lesion and sacrificing the mental nerve



Fig 7: Intra oral site post 6 months with uneventful occurrence

Fig 7: facial profile of the patient with no signs of recurrence

III. Discussion:

Neurothekeoma, a rare benign tumor of the nerve sheath, was initially described by Harkin and Reed, in 1969. The term Neurothekeoma was described by Gallagher and Helwing, in the 1980s.³ It is commonly present as small solitary nodules on the face and the upper limb; however, other anatomical sites, such as oral cavity, cauda equina, shoulder and neck have been reported, where in oral involvement is extremely rare.^{4,5}

Histopathologically, these lesions reveal circumscribed tumor mass composed of epithelioid and spindle cells, arranged in well-formed micronodules.⁶ The basic treatment for neurothekeoma is complete excision of the lesion with clear margins. No malignant transformation or metastases have been reported and local recurrence is extremely uncommon, but possible, especially for the cellular/mixed group. Recurrence of the same is usually due to incomplete excision.^{7,8}

The differential diagnosis of neurothekeoma may include other neural entities, such as schwannoma, true neuroma and myxoid neurofibroma.⁹ They can be distinguished by clinical presentation, histological features, and immunohistochemical stains.

The immunohistochemical markers for neurothekeoma are NKI/C3(+ve), vimentin(+ve), NSE(+ve), HMB45(-ve), S-100 (-ve).¹⁰

Clinical presentation of neurothekeomas are asymptomatic slow-growing lesions, that are usually benign, solitary and dome-shaped lesions that rarely ulcerates the skin.⁹

Clinicians should consider the possibility of an early local recurrence, thus must follow a close clinical follow-up.

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