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Sublingual Nonsendromik Soliter Nörofibrom Olgu Sunumu

A Case Report of Sublingual Nonsyndromic Solitary Neurofibroma

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Öz

Giriş: Nörofibrom periferik sinir kılıfından köken alan, yavaş büyüyen, benign bir tümördür. Baş ve boyuna yerleşik nörofibromların yüzde 6'sı oral kavitede görülmektedir. Oral kavitede ağız tabanına yerleşik soliter nörofibrom oldukça az görülmektedir. Bu olgu sunumunun amacı bu bölgede nadir görülen bu tümörün tanı ve tedavisine dikkat çekmektir.

Olgu Sunumu: Otuz iki yaşında erkek hasta kliniğimize dil altında şişlik şikayetiyle başvurdu. Fizik muayenede ağız tabanında, dili yukarı iten, yaklaşık 3 cm büyüklükte üzeri normal mukoza ile kaplı kitle mevcuttu. Palpasyonda ağız tabanından submandibular bölgeye uzanan hareketli, ağrısız ve sert bir kitle saptandı. Hastaya cerrahi tedavi önerildi. Genel anestezi uygulanarak kitle intraoral yolla eksize edildi. İmmünhistokimyasal incelemede tümör hücrelerinde S-100 ile pozitif immünreaktivite izlenmesi nedeniyle nörofibrom tanısı kesinleştirildi.

Sonuç: Ağız tabanında şişlik şikayeti ile başvuran hastada ön tanılar arasında nörofibromun da yer alması gerektiği, özellikle malign dönüşüm riski taşıyan bu tümörün sendromik hastalıklarla ilişkili olabileceği akılda tutulmalıdır.

Anahtar Kelimeler: Sublingual, Nörofibrom, Nörofibromatozis

Abstract

Introduction: Neurofibroma is a slowly growing, benign tumor originating from the peripheral nerve sheath. Neurofibromas observed in oral cavity constitute 6% of those located in head and neck region. Solitary neurofibroma located in the flow of the mouth in oral cavity is observed quite low. The purpose of this case report is to attract attention to the diagnosis and treatment of this rare tumor located in this region.

Case Presentation: Thirty-two years old male patient applied to our clinic with swelling under his tongue. In physical examination, a mucosa covered mass was determined that is approximately 3 cm in size and pushing the tongue upwards. In the examination with bimanual palpation, mobile, painless and hard mass was observed to extend from the floor of the mouth to submandibular region. Surgical treatment was recommended to the patient. The excision was performed under general anesthesia and through intraoral route. Immunohistochemical examination confirmed the diagnosis of neurofibroma due to positive immunoreactivity with S-100 in tumor

cells.
Conclusion: It is important to consider neurofibroma among pre-diagnoses for patients who have applied with swelling on the floor of their mouth, and especially to recognize that this tumor, which has a malignant transformation risk, may be associated with syndromic diseases.

Introduction

Neurofibroma is a slow-growing, benign, nerve sheath tumor that originates from peripheral nervous system. It is observed as 20% among benign peripheral nerve sheath tumors. Neurofibroma located in head and neck constitute approximately 25% of all neurofibromas and 6% of those are observed in oral cavity. Involvement of tongue, hard and soft palate, gingiva, mandibula, tongue base and rarely floor of the mouth are observed in oral cavity (1).
World Health Organization (WHO) has grouped neurofibroma under three groups; localized, diffuse and plexiform (2). The localized neurofibroma is the most common, and occurs sporadically in the majority of cases. They are generally detected between the age 20-30 and show an intraneural, non-infiltrative growth. Diffuse neurofibroma generally presents in children and adolescents as plaque-like blisters on the skin of head and face. Generally, they are not syndromic. Plexiform neurofibroma is the most rarely observed one and mostly originates from distal end of peripheral nerves. It is often associated with neurofibromatosis type 1 (NF1) and multiple endocrine neoplasia type 2B (MEN-2B). NF1 can be determined in the physical examination in approximately 27% of patients who have plexiform neurofibroma (3).
Solitary neurofibroma located in the flow of the mouth in oral cavity is observed quite low, and the purpose of this case report is to attract attention to the diagnosis and treatment of this rare tumor.

Case Presentation

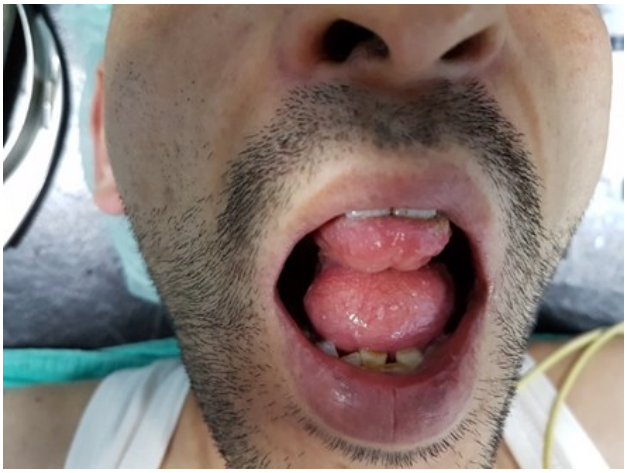

Figure 1: The mass caused difficulties in speaking and eating by pushing the tongue towards superior

Thirty-two years old male patient applied to our clinic with swelling under his tongue. Present complaint of the patient has been persisting for approximately one year, however he has applied to our clinic since he had been having difficulties in speaking and eating in the last month. In physical examination, a mucosa covered mass was determined that was approximately 3 cm in size and pushing the tongue upwards. Both wharton ducts were open (Figure 1). In the examination with bimanual palpation, mobile, painless and hard mass was observed extending from the floor of the mouth to the submandibular region. Tongue movements were observed to be limited due to the effect of the No loss of feeling was described for taste. His both commissures of

lips were observed to be symmetrical and no limitation was observed in lips and tongue movements. No skin or skeletal pathology were determined in the systemic examination that suggests neurofibromatosis or multiple endocrine neoplasia syndrome.

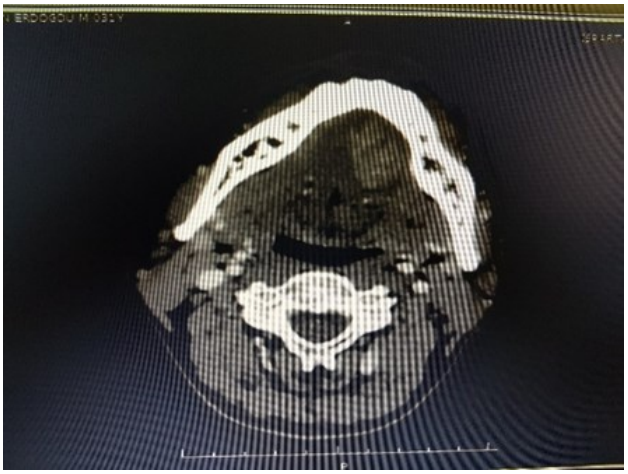

Figure 2: Well-defined mass non-invasive to mandibula and adjacent tissues with hyper and hypointense foci

The result of fine-needle aspiration biopsy, that had been performed previously in an external center, was reported as 'blood elements' and a computed tomography had been performed (Figure 2). Preoperative laboratory tests were normal for the patient. There were no additional characteristics for his medical history or familial history. Surgical treatment was recommended to the patient. The excision was decided to be performed under general anesthesia and through intraoral route since the mass was smaller than 6 cm, similar to a dermoid cyst located on the floor of the mouth (4). The mass was reached by performing a horizontal incision on the floor of the mouth far from both wharton ducts. The mass was dissected from surrounding tissues. During dissection, both wharton ducts and lingual nerve were protected, and no adhesion or fistule tract was observed associating with the mass. After the mass has been removed with no residues, bleeding control was performed and operation was terminated (Figure 3, 4).

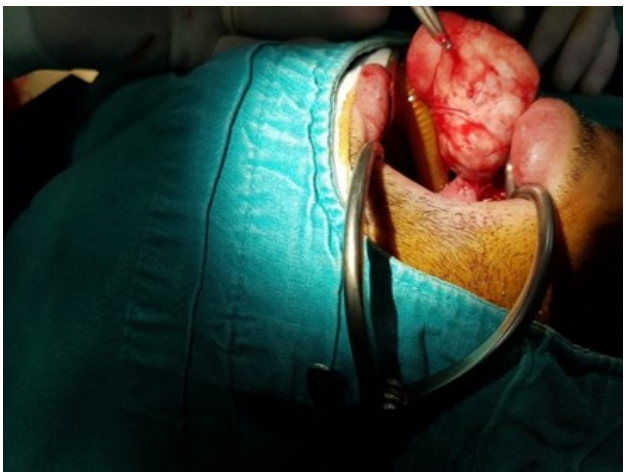

Figure 3: Intraoral excision of the mass

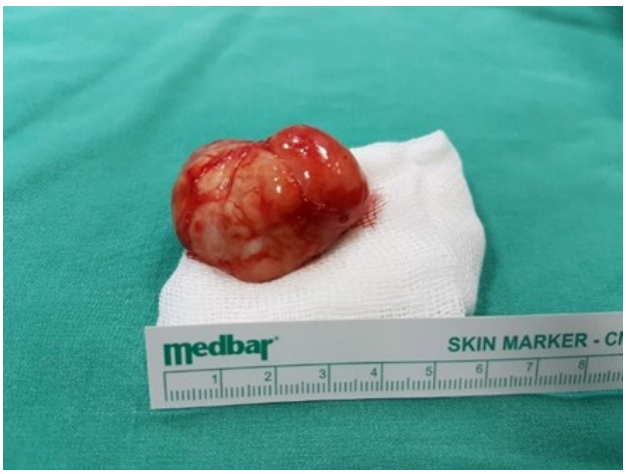

Figure 4: Macroscopic appearance of the specimen

An immunohistochemical study was performed after observing spindle cells on light microscopy in the histological examination, and diagnosis of neurofibroma was finalized since they were observed to be S-100 (+). (Figure 5,6) At the end of the first week, normal saliva secretion was observed from wharton duct.

Tongue movements were normal inside and outside of the mouth in the post-operative control and there were no complaints of impaired taste. Patient had no additional problems. No findings suggesting neurofibromatosis or multiple endocrine neoplasia syndrome were determined in the repeated systemic examination. There were no cafe au lait spots on the body and family history for neurofibromatosis was negative.

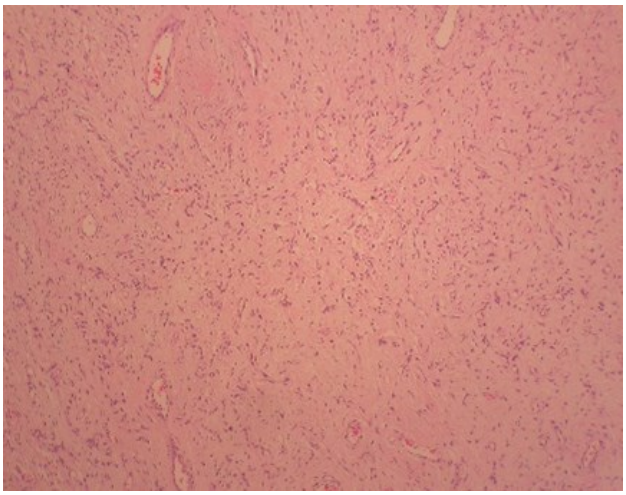

Figure 5: Tumor composed of spindle cells (HE x100)

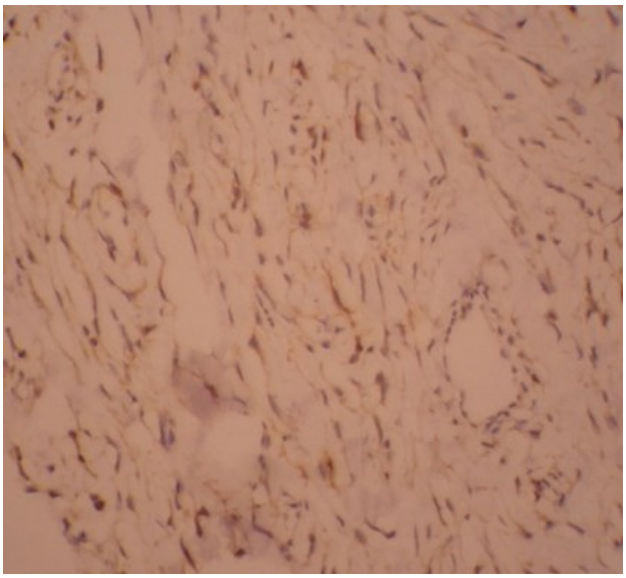

Figure 6: S-100 cytoplasmic positivity (DABx400)

Discussion

Although neurofibromas are benign, rapid growth of the existing mass for long periods may suggest possible malignant transformation. Malignant transformation prevalence of neurofibroma is reported as 2%-29%. (3). The malignant transformation risk of plexiform neurofibroma accompanying syndromic disease is high and it is assessed as the precursor of malignant peripheral nerve sheath tumor however, malignant transformation of localized solitary type neurofibroma has been reported (5 -8).

NF1 disease is generally diagnosed by clinical determination of classic triad and finalized with genetic analysis. This triad consists of cutaneous lesions, mental retardancy and skeletal anomalies. The presence of multiple neurofibroma is important in NF1 diagnosis, but non-syndromic multiple neurofibroma is observed rarely (9). For the pre-diagnosis of neurofibroma, the presence of cafe au lait spots should be sought in patient's general medical examination and a detailed history should be obtained from the patient. Upon determining the presence of accompanying skeletal anomalies, mucosal multiple neurofibroma in physical examination, the patient should be directed to genetic analysis with the pre-diagnosis of multiple endocrine neoplasia.

Etiology of solitary neurofibroma has not been finalized. The tumor is considered to originate from schwann cells or perineural fibroblasts (10). Some authors suggest that hamartomatosis or simple hyperplastic process may play a role in the etiology and development is not due to neoplastic process (11). Papadopoulos et al. (12) stated that neurofibroma located on the floor of the mouth may be developed from fifth and seventh cranial nerve fibers. In our case, no findings suggesting paresis\paresthesia of fifth or seventh cranial nerve were determined in postoperative period. Also, it was observed in the literature review performed for the etiology of neurofibroma that, a case has developed following radiotherapy isolated in the floor of

the mouth (13).

Macroscopic appearance of localized neurofibroma varies according to extraneural or intraneural growth. Cutaneous localized neurofibroma generally goes beyond perineurium, it is unencapsulated but it is a well -defined mass. When deep soft tissue localized neurofibroma shows intraneural growth, it is generally a well-defined and capsulated mass similar to schwannoma. It can present as white colored mass on the floor of the mouth, feeling hard with palpation (2). We present 32 years old man who had a well defined and capsulated mass at sublingual region. He was not telling about rapid growth of the mass but he had swallowing difficulties. In his general medical examination there was not any café au-lait spots or accompanying skeleton anomalies. The mass was pink-white colored and feeling hard with palpation.

Through a histological aspect, localized neurofibroma consists of interpenetrating spindle cells that are long and thin. Unlike schwannoma, it does not contain Antoni-A and B regions, myxoid regions and degenerative regions are less. In the immunohistochemical analysis, positive S-100 protein finalized light microscopy diagnosis (2).

In the differential diagnosis of solitary neurofibroma, dermoid cyst, branchial cleft cyst, thyroglossal cyst, ectopic thyroid tissue should be considered for developmental aspect, sublingual abscess, cellulitis, Ludwig angina for infectious aspect, sialoadenitis, mucocele, ranula for salivary gland aspect, cystic hygroma for hamartomatosis, squamous cell carcinoma for neoplasia; lipoma, lymphangioma, epidermoid, lymphoepithelial, heterotypical gastrointestinal and enteric duplication cyst should be considered.

The treatment of peripheral nerve sheath tumors is surgery. In the presented case, being solid mass in radiological examinations, surgical excision has been performed with benign peripheral nerve sheath tumor pre-diagnosis. During surgery, care should be taken for protecting the integrity of lingual nerve and Wharton ducts. Relapse rate is very low for benign peripheral nerve sheath tumors in the long term follow-up. Intraneural or extraneural growth of the tumor may change treatment options. Complete recovery is obtained and relapses are rare in well-defined and localized tumors, as in our case. However, in patients with extraneural or infiltrative growth, sacrificing the nerve may be required for complete excision. If protection of the nerve is important, debulking or medical therapy may be tried. In medical therapy, treatment interventions performed with oncolytic viruses, phase 3 trials of which are completed successfully, seem promising (14).

Isolated solitary neurofibroma in the floor of the mouth is observed rarely. Al-Omran et al. (15) have reported an isolated solitary neurofibroma that has developed from the floor of the mouth in 2006 and indicated as a result of their Medline search that there was no previously reported case. Afterwards, in 2011 Maruyama et al. (16) have presented a case with similar characteristics. As a result of their search, they reported 66 neurofibromas developed from facial area, and they have found a previous case in 1970s which was the first report of neurofibroma originating from the floor of the mouth, however there were no detailed descriptions.

In conclusion, we report a very rare case of solitary neurofibroma arising in the floor of the mouth. Immunohistochemistry supports the pathological diagnosis of this case.

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