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CASE REPORT

EXTRA-OSSEOUS SOLITARY HARD PALATE NEUROFIBROMA: A CASE REPORT

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ABSTRACT:
Neurofibromatosis (NF) is a term that has been applied to a variety of related syndromes, characterized by neuroectodermal tumors arising within multiple organs and autosomal-dominant inheritance. Neurofibroma may occur as solitary lesion or as part of a generalised syndrome of neurofibromatosis or very rarely as multiple neurofibromas without any associated syndrome. There are two distinct variants of neurofibromatosis type I and type II. We present a case of neurofibroma of the hard palate associated with neurofibromatosis type I. Diagnosis was made on the basis of history, clinical findings and histopathology. Literature was reviewed and incidence and frequency of different cases reported in oral cavity including treatment and recurrence are discussed.
Keywords: Neurofibromatosis type-1, Neurofibroma, Benign tumor, Hard Palate

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INTRODUCTION

The neurofibroma (NF) is a benign tumor of the peripheral nerve sheath that rarely affects the head and neck. However, among neural lesions, this is the one that most frequently affects this region.[1,2] The NF can be intra or extra-osseous, alone or multiple (associated with type I neurofibromatosis).[2,3] The most common extra-osseous mouth NF locations are tongue, oral mucosa and lips[2]. In the literature we found two well-documented cases of solitary extra-osseous neurofibroma in the hard palate.[1,4]

CASE REPORT

A 37 year-old-male was referred to us because of a diagnosis of a lesion on the palate. The patient had a bilobed, asymptomatic, sessile and fibrous pink and smooth nodule measuring 35 x 25 x 05 mm, on the left side of the posterior region of the hard palate, near the alveolar border, that had been evolving for four years. His medical history was uneventful. X ray images did not show any alterations. The clinical diagnosis was pleomorphic adenoma or benign mensenchymal neoplasia. We did an incisional biopsy and the specimen was referred to analysis. Histological exam showed fusiform cell proliferation with undulated nuclei distributed in a disorganized fashion on the fibrous connective tissue.

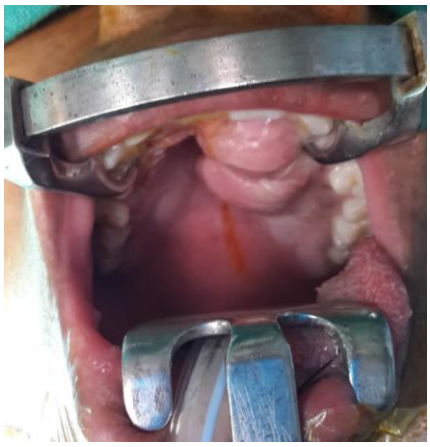

Figure 1: Lobulated swelling on palate

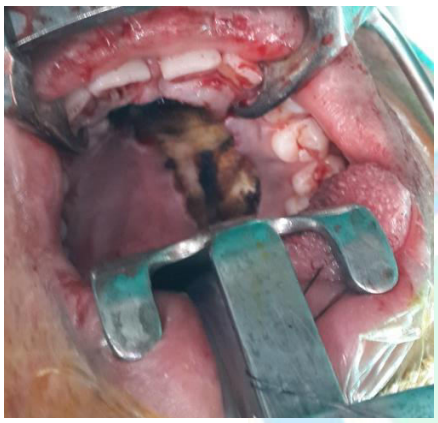

Figure 2: Surgical excision of lesion

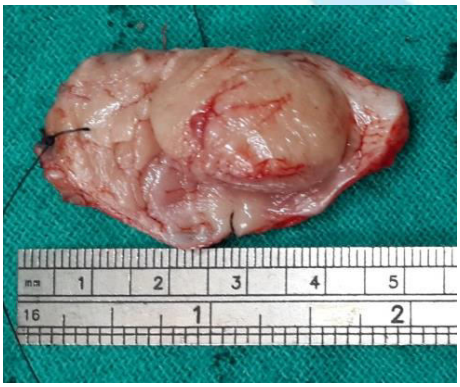

Figure 3: Gross specimen

All the neoplastic cells were immunopositive for protein S-100 (streptoavidin-biotin technique), Dako Corporation®, clone: Z0311, dilution 1:100, without antigenic recovery, incubated for 18 hours at 4ºC) (Figure 1C).

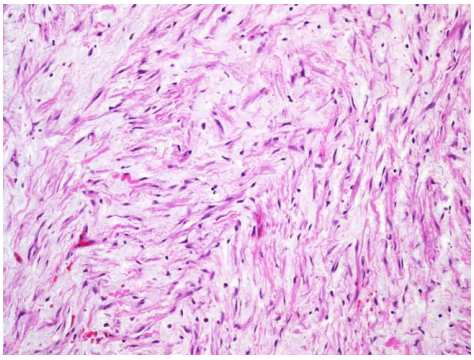

Figure 4: Spindle shaped fibroblasts and nerve cells

The final diagnosis was neurofibroma. The patient was re-evaluated and there were no more evidences of type I neurofibromatosis. The lesion was excised; it was well outlined and attached to the greater palatine nerve (Figure 1-4). This portion of the nerve was also resected (Figure 1E). There was no recurrence during the 15 months of follow up.

DISCUSSION

Pollack [1] and Shimoyama et al[4] reported two cases of solitary extra-osseous hard palate neurofibroma, which usually is a small, sessile, smooth, well outlined and non capsulated nodule.[1,2] Cherrick and Eversole[2] observed a predilection for females. Chen and Miller[5] reported that mouth neurofibroma affect people between 9 and 72 years of age. These clinical characteristics were seen in this case. neurofibromas are immunopositive for the S-100 protein in 85 to 100% of the cases, indicating its neural origin.[3,6] Treatment for solitary NF is surgical excision and recurrence is rare.[2,4] In the case hereby described the tumor was easily removed because it was well outlined. Moreover, a portion of the greater palatine nerve was also removed.

CONCLUSION:

It is fundamental to follow the patient with neurofibroma, because the solitary neurofibroma can be the first manifestation of type I neurofibromatosis. This patient is under follow up care and until this report was made we did not see any relapse.

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