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Solitary Neurofibroma of the Lip: Report of a rare case

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ABSTRACT

Background and Aim: Neurofibroma is an uncommon lesion in the oral cavity. Most reported cases are multiple and associated with the generalized neurofibromatosis syndrome. Rarely, neurofibroma is formed as a solitary lesion and is not associated with the syndrome. To our knowledge, only four cases of solitary neurofibroma have been reported in the literature, which makes the clinical diagnosis difficult.

Case Presentation: We report a case of solitary oral neurofibroma in the lower labial mucosa of a 43-year-old woman. These patients need regular follow-ups in order to identify recurrences and probable ancillary features of neurofibromatosis syndrome which may be life-threatening to the patient.

Conclusion: Solitary neurofibroma should be considered in the differential diagnosis of the Intra oral nodules beside more common lesions including fibroma, mucocele and pleomorphic adenoma.

Case Report

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Introduction

Peripheral Nerve Sheath Tumors are uncommon in the oral cavity. This group of lesions are seen in the peripheral nervous system and are divided to two categories of benign and malignant tumors. Benign tumors include Neurofibroma and Schwannoma which arise from perineural fibroblasts and/or Schwan cells. The malignant tumor of this group of lesions is Malignant Peripheral Nerve Sheath Tumor which either originates primarily from peripheral nerve sheath cells or occurs secondarily to the benign tumors of peripheral nerve sheath. [1-4]

Although Neurofibroma is the most common peripheral nerve tumor, it is rarely seen in the oral cavity. Most cases of Neurofibrom in the oral cavity are multiple (three lesions or more) and have been reported as a part of generalized neurofibromatosis syndrome. But rarely, this lesion can be solitary (one or two lesions) with no visceral manifestations. To the best of our knowledge, to date only four cases of solitary Neurofibroma have been reported in the lip.[4-8]

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Skin is the most common area involved by Neurofibroma and this lesion usually forms in the third decade of life. [1-3] Pathogenesis of solitary Neurofibroma is unclear but neurofibromatosis is an autosomal dominant disease with at least 8 known types. The most common type of neurofibromatosis is neurofibromatosis type 1 (NF1) or Von Recklinghausen Disease (VRD) which comprises 85 to 97% of the cases. The manifestations of this disease are highly variable. In some patients, only a few number of Neurofibromas are present while in some others hundreds to thousands of Neurofibromas can be detected. [8-10]

In the present study, we report a case of solitary oral Neurofibroma in the lower labial mucosa of a 43-year-old woman. This patient showed no symptoms or familial history of neurofibromatosis syndrome and the diagnosis of solitary Neurofibroma was confirmed in this case.

Case Presentation

A 43-year-old female with a chief complaint of lip swelling referred to the dental school of Tabriz University of Medical Sciences in 2012. In her medical history no systemic, endocrine or metabolic disorder was mentioned. Extra oral examination revealed normal results. In intra oral examination, a pale pink, soft and well-defined nodule with approximate dimensions of 7×5×5 mm was seen on the lower right labial mucosa of the patient. (Figure 1)

Figure 1- Clinical appearance: pink, soft, well-defined solitary nodule is seen on the lower labial mucosa of the patient

The superficial mucosa was stretched, the surface of the lesion was glossy without any signs of hemorrhage, pulsation or suppuration. The patient mentioned that the lesion had caused no pain, burning sensation or sensitivity, had a gradual growth and has reached this size in 6 months. The differential diagnoses were Fibroma and Neurofibroma and excisional biopsy was performed for diagnosis confirmation. In histopathological evaluation, an un-encapsulated subepithelial tumoral growth was seen. Interlaced bundles of spindle-shaped cells with elongated and dark, ovoid or wavy nuclei with irregular arrangement were seen in a loose connective tissue matrix. Variable amounts of delicate collagen fibers and mast cells were present dispersedly in the lesion. (Figure2)

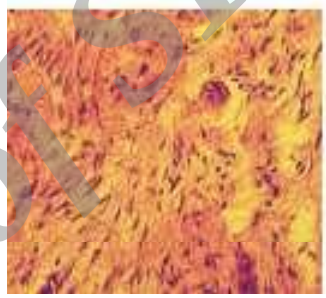

Figure 2- Histopathological appearance: spindle-shaped cells with ovoid and wavy nuclei along mast cells in a loose connective tissue matrix (H&E staining, 400× magnification)

In immunohistochemical analysis, positive reaction to S-100 protein was detected which proved the neural origin, (Figure 3) and histology and immunohistochemistry findings allowed the diagnosis of Neurofibroma.

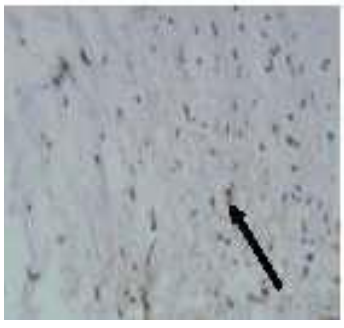

Figure 3- Histopathological appearance: expression of S-100 protein is seen in some cells. (immunohistochemical staining, 400× magnification)

The syndrome manifestation was rejected as the patient showed no sign of neurofibromatosis disease, no serious anomaly was detected on radiographic evaluations and no familial involvement by this syndrome was present and diagnosis of solitary Neurofibroma was confirmed. Two years after the surgery, the patient was still non-symptomatic without any signs of recurrence or neurofibromatosis syndrome features.

Discussion

Neurofibromas are usually non-symptomatic, grow slowly and vary in size from small nodules to large masses. (2-3) the present patient also had a relatively small, non-symptomatic and slow growing lesion. The other four previously reported cases also had a non-symptomatic lesion with slow growth. Two cases were large masses and two others were small nodules relatively similar to the clinical characteristics of the present lesion. None of the reported cases had signs of systemic disease or history of trauma in the area of the lesion.

From the four reported cases, three cases were reported in 10 to 20 year-olds and one was seen in the age of 60. Our patient was 43 years old. Considering all these five cases, it seems that although oral solitary Neurofibroma is seen over a wide age range but the probability of involvement is higher in young adults (the second decade). From these five cases, oral solitary Neurofibroma was detected in 3 females and 2 males with three cases in the lower lip and 2 cases in the upper lip. It is probable that this lesion is more prevalent in females and in the lower lip.

The treatment method for solitary Neurofibroma is surgical local excision and malignant transformation may be seen in some cases. If the lesion cannot be separated from the originating nerve, the related nerve may be amputated. Multiple Neurofibromas in Neurofibromatosis syndrome may be excised due to functional, respiratory or cosmetic disturbances but this procedure is difficult when they are numerous. Neurofibromatosis treatment is mostly directed towards the prevention and problem management for the patient. One of the most serious complications of this disease is malignant transformation (mostly malignant peripheral nerve sheath tumor) with poor prognosis. CNS tumors, leukemia, pheochromocytoma, rhabdomyosarcoma and wilms' tumor may also be detected in these patients. These patients need regular follow ups in order to be treated on occurrence of the primary signs of malignancy. (11-13) The present patient responded well to surgical excision and showed no signs of recurrence or Neurofibromatosis syndrome features in the 2-year follow up. The other four patients had also responded well to surgical excision and no signs of recurrence were detected after maximum of 18 months of follow up.

Conclusion

Solitary neurofibroma should be considered in the differential diagnosis of the Intra oral nodules beside more common lesions including fibroma, mucocele and pleomorphic adenoma.

Conflict of interests

Authors report no conflict of interest related to this study.

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