



Solitary central neurofibroma of the maxilla: A Case Report

Neurofibroma central de maxilla: relato de caso

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ABSTRACT

Neurofibroma is a benign tumor which rarely occurs in the oral cavity. Generally, oral neurofibroma is part of a neurofibromatosis type-1 like syndrome (Von Recklinghausen's disease) and rarely occurs as a solitary lesion of oral cavity. Up to now, few cases have been reported with the solitary neurofibromas of tongue and submandibular gland. The tumor can also be rarely located centrally in the bone. This research presents a central neurofibroma case in the maxillary bone which has no relationship with the neurofibromatosis.

KEYWORDS

Central neurofibroma; Neurofibromatosis; Solitary neurofibroma; Von Recklinghausen's disease.

RESUMO

O neurofibroma é um tumor benigno que raramente ocorre na cavidade oral. Geralmente, oral neurofibroma é parte de da neurofibromatose tipo 1 (Von Recklinghausen's doença) e raramente ocorre como uma lesão solitária da cavidade oral. Até agora, poucos casos foram relatado com os neurofibromas solitários de língua e glândula submandibular. O tumor também raramente pode estar localizado centralmente no osso. Esta pesquisa apresenta um neurofibroma central caso no osso maxilar que não tem relação com a neurofibromatose.

PALAVRAS-CHAVE

Neurofibroma central; Neurofibromatose; Neurofibroma solitário; De Von Recklinghausen doença.

INTRODUCTION

Neurofibroma is a rare benign tumor of peripheral nerve sheath. In most of the cases, it is almost circumscribed, but not capsulated. Solitary lesions also happen rarely. [1,2].

In a series studied by Peterson *et al.* (1932-1952), the intra oral neurofibromas occurrence was 6% in those patients with neurofibromatosis. [3] In another research on 19 neurofibromatosis cases studied by cherrick and Eversole (1971), it was indicated that 20% of patients with neurofibromatosis had intraoral neurofibroma. [4] In 1980, Wright and Jackson reported that the relationship between the neurofibromas and von Recklinghausen' disease is about 60% [2].

Neurofibromas are soft and doughy exophytic masses which have slow growing characteristics without any pain. In 1969, Gupta *et al.* found that 9% of the 303 cases of benign nerve sheath tumors occurred in the oral cavity [5]. It has been reported that the most common intraoral sites of neurofibroma are the tongue and buccal mucosa. [6]

Neurofibromas of palate, gingiva, and intraosseous varieties have been reported in mandible and maxilla. [7,8] Solitary intraosseous neurofibroma is a rare benign tumor with very few reported cases in the literature. [6] This case report presents a rare solitary central neurofibroma case in the maxillary bone which has no relationship with the neurofibromatosis.

CASE REPORT

A 3 y/o male child had come to our center with an asymptomatic enlarging swelling in his posterior right nasomaxillary region, since 11 months ago. An operation was done for him once before by incisional access with the fibrous dysplasia diagnosis approximately 5 months ago, but after that the condition became worse and the patient came to our center in oral and maxillofacial medicine center of Shiraz University of medical science in order to find a treatment. Based on the patient's familial background, the patient himself and his first relatives had no systemic disease history or the same swelling in the past.

In the extra oral examination, the swelling was obvious (Figure 1). Intraorally solitary swelling was also observed extending buccally from maxillary lateral incisors to the tuberosity (Figure 2).



Figure 1 - Extra oral swelling.



Figure 2 - Intraoral swelling.

The lesion surface was smooth and had a normal pink-color, similar to the normal mucosa.

The following differential diagnosis was considered after the clinical examination:

- 1) Fibrous dysplasia
- 2) Giant cell lesion

Radiological examination was accomplished. Computed tomography (CT) of face presented an expansile calcified lesion measuring 55, 48, and 46mm in the right maxillary sinus with its extension to the right side of the face soft tissue, and also to the right ethmoidal air cells, due to the thin lamina papyrus. Extension to the right nasolacrimal duct with pressure effect on the right orbit inferior wall was obvious, which caused exophthalmos, also along with the mild nasal septal deviation to the left. Right maxillary alveolar ridge involvement was obvious which caused few floating teeth (Figure 3).

An incisional biopsy was performed again, and this time the result showed central neurofibroma (Figure 4).

As we mentioned earlier, histopathological examination of the former biopsy indicated the fibrous dysplasia, but after the second biopsy. Immunohistochemically (IHC) research reported the spindle tumoral cells in a myxoid background, which are positive for S100 diffusely, and also negative for the CD34 and EMA, and proliferative index (Ki67) is about 3%, confirming that the benign spindle cell neoplasm with neural origin were consistent with the neurofibroma.

The lesion was completely excised after the diagnosis with IHC establishing (Figures 5 and 6). Hemimaxillectomy and reconstruction of the right orbital floor and the right lateral wall of the nose were accomplished under the general anesthesia by an oral and maxillofacial surgeon specialist (Figure 7).

Although, the tumor course assumed to be aggressive, but in histological sections, mitosis was less than 4/10 HPF, and also S100 was diffusely positive, as a result malignancy was ruled out.

On the 30-month follow-up, the results were satisfactory (Figure 8).

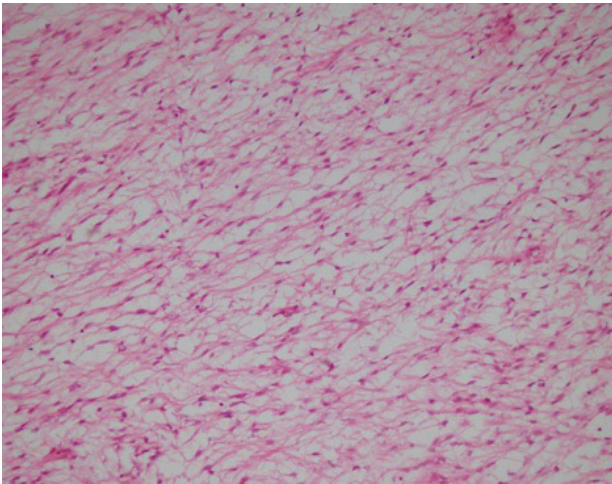


Figure 3 - A. CT (axial plane) revealing the expansile calcified lesion causing bone destruction in the right maxillary sinus with its extension to the right side of the face soft tissue, and also to the right ethmoidal air cells, due to the thin lamina papyrus. Extension to the right nasolacrimal duct with pressure effect on the right orbit inferior wall was obvious; B. CT (coronal plane)



Figure 4 - H&E stain showing the spindle tumoral cells in a myxoid background.

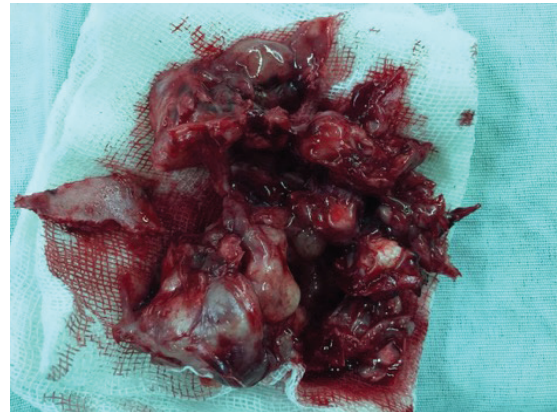


Figure 5 - ???.

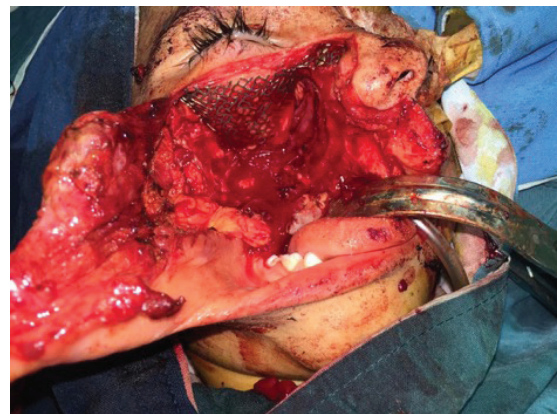


Figure 6 - Removed tissues during the surgery.



Figure 7 - A. Reconstruction procedure of the lower orbital rim and lateral wall of the nose during surgery; B. The patient's profile 10 days after surgery

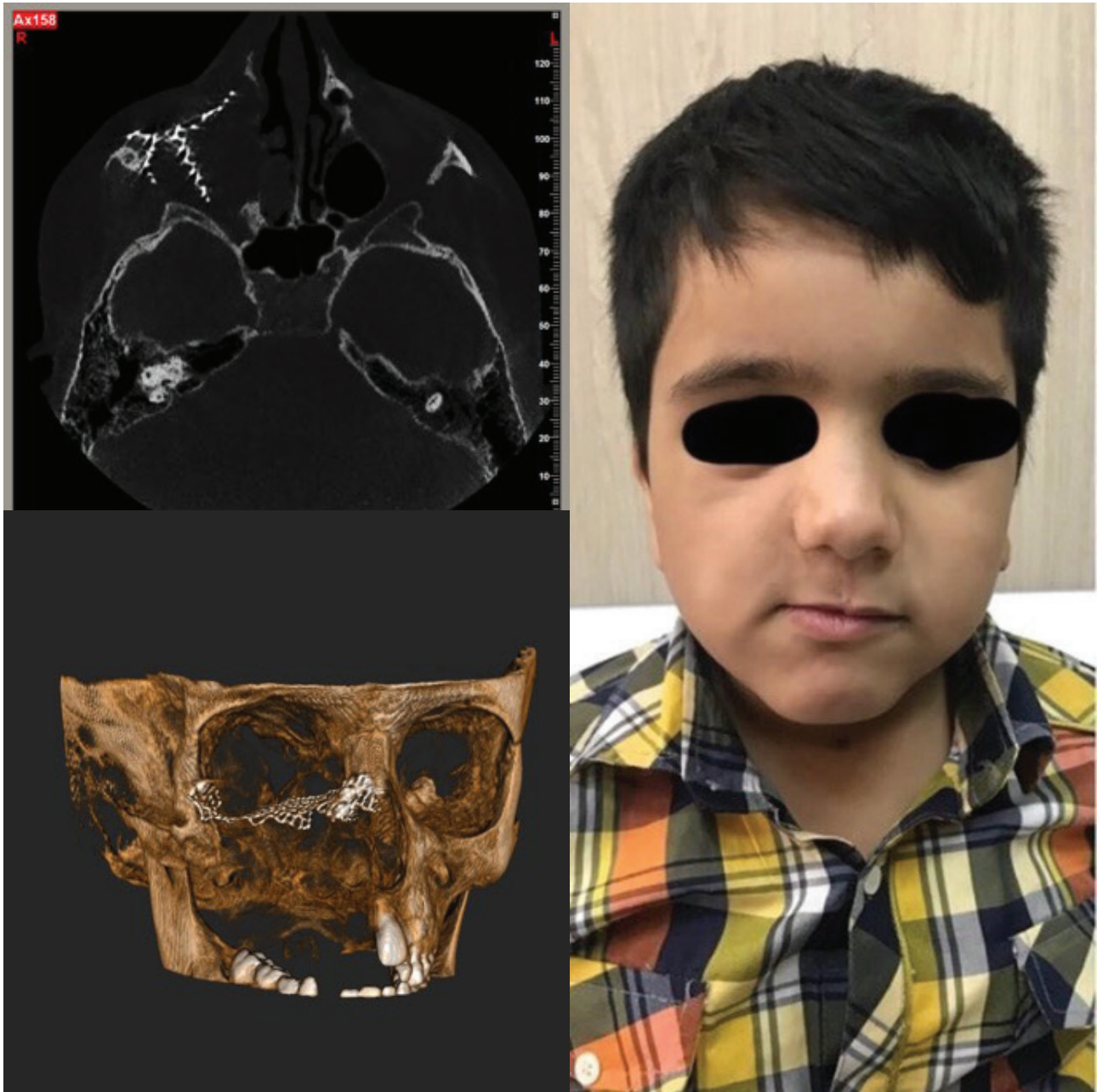


Figure 8 - A. CBCT views on the 30th Month follow up; B. CBCT views on the 30th Month follow up; C. 3D image on the 30th month follow up; D. The patient's profile on the 30th month follow up

DISCUSSION

Neurofibromas are slowly growing benign tumors. The neurofibroma origin is considered to be perineural fibroblasts, which originates from neuroectodermal tissue. [9]

Neurofibromatosis was described in 1882 by von Recklinghausen at first, and neurofibromatosis

type 1 is its most common type. It is a syndrome with multiple neurofibromas, café-au-lait spots, and some bony changes like macrocephaly, pseudo arthrosis especially tibia, cerebral kyphosis, and bowing. [10] The individual with at least two of the followings criteria is identified as neurofibromatosis type 1:

1) 6 or more Cafe-au-lait spots, which are light brown well circumscribed macules with regular border that should be greater than 5mm in Youngers and more than 15mm in adults.

2) At least two neurofibromas of any form, or one plexiform neurofibroma

3) Crow's sign, which are axillary and/or inguinal freckles

4) Optic glioma

5) at least Two Lisch nodules (iris hamartomas)

6) Bone lesions like sphenoid dysplasia or thinning of the long bones' cortex with or without pseudo arthritis

7) Affliction of one of the first relatives [11]

NF1 is genetically different from type 2, and is the most common autosomal dominant inherited disorder with an incidence rate of 1 in 3000 [12]. NF1 gene is positioned on chromosome no. 17 and its mutation leads to neurofibromin protein absence encoded by this gene, and also other severe developmental abnormalities, because in many tissues, neurofibromin's role is as same as GTPase activator. [9]

There are two types of neurofibromas clinically, subcutaneous and Plexiform variety. As subcutaneous variety improves at peripheral nerve endings, it is going to be identified also as localized neurofibroma. It appears in late childhood or early adolescence, generally, and contains about 95% of cases. Plexiform variety develops in the body deeper areas near to the nerve roots, and is approximately consisted of 5% of cases. It generally appears during the first 2 years of life. Plexiform neurofibromas have the potential of malignant transformation to malignant peripheral nerve sheath tumors (MPNST). [12]

Neurofibromas occur generally as the multiple lesions, and are usually in relationship with the NF1, solitary tumor, which rarely happens in our case. As there was no occurrence history of such lesion in the family, this is a sporadic variety case. Most of the intra osseous lesions reported cases are in the posterior mandible, and a few cases are in maxilla. [6,13] The thick nerve bundles of

inferior alveolar nerve are its occurrence in the mandible predisposing factor. [9,13] It happens twice in females. [9] Its occurrence is between the age of 10 months and 70 years old, but it is common mostly during the third decade of life. [14]

In a case reported by Dalili et al. (2012) there was mandible, maxilla, and orbit concurrent involvement. Radiographically, the lesion showed bone destruction in the maxilla, and the walls of right maxillary sinus displacement into the antral cavity. Multiple impacted maxillary molars were also observed. [13] Similar extensive bone destruction and the nasal cavity displacement floor were presented along with the maxillary sinus walls. In the mandible, neurofibroma might show features like foramen and inferior alveolar canal enlargement, deep sigmoid notch, condyle deformity, unerupted teeth, and a cyst like lesion. [13] In the present case, the lesion occurred in the right maxillary sinus with its extension to the right side of the facial soft tissue and also to the right ethmoidal air cells, due to the thin lamina papyrus. Extension to the right nasolacrimal duct with a pressure effect on the right orbit inferior wall was obvious which caused exophthalmos along with a mild nasal septal deviation to the left. Right maxillary alveolar ridge involvement was obvious which caused limited floating teeth.

Another study on the solitary neurofibroma was reported by Narwal et al. in 2008 with a 5-month-old child which involved maxillary alveolar. [9] It extended from the right maxillary alveolar region up to the right lateral surface of the nose, measured 2cm × 1cm, radiographically. Our case, measured at 55, 48, and 46mm, indicated similar performance along with the sinus displacement.

A case of neurofibroma of palate was reported by Sreenivasa et al. in 2014 and no bony involvement was observed on panoramic and PA skull view surprisingly. [15]

Although intraosseous neurofibromas are slow growing benign tumors which cause surrounding bones less destruction, it showed a rapid growth over a period of 15 months in our case, with large bone destruction in the maxilla.

Solitary neurofibromas and the lesions observed in the neurofibromatosis have the same histopathological features. [2] These tumors are not capsulated and are a mixture of Schwann cells, perineural cells, and endoneural fibroblasts. [16,17] A mixture of nerve fibers on an irregular distributed collagen matrix, a few spindle cells, and mucinous stroma can be observed. [10] Mast cells and hyaline changes might be present in older lesions frequently. The cells are uniformly positive for s100protein, due to their neural crest origin. [1]

The gold standard treatment of solitary neurofibroma is complete surgical excision; similar to the one we performed for our case. [14] Radiotherapy alone is not adequate, but it can reduce the tumor size and growth. [14,18] Diode lasers presented good results for small and accessible tumors. [18]

When multiple neurofibromas are associated with neurofibromatosis, neurofibrosarcoma can happen for approximately 5-6% of the cases. [2] Pheochromocytoma is also observed in these cases more than others. The malignancy changes risk in solitary neurofibroma is unidentified, yet. [2]

CONCLUSION

Intraoral neurofibroma is an uncommon pathology which rarely occurs centrally and, in a condition of central occurrence, it takes place in mandible in most of the cases. Consequently, this case of solitary central neurofibroma rarely happens in the nasomaxillary region.

“Conflict of Interest: None”

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