Neurofibroma of the maxillary antrum: A rare case

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Abstract

Neurofibromas are benign tumors of peripheral nerve tissue, frequently associated with neurofibromatosis type 1. Their isolated occurrence in the maxillary antrum is rare, with only 6 cases described in the English literature to the best of our knowledge. Primary neurogenic tumors in the maxillary sinus are unusual entities. The majority of the reported cases that have dealt with neurilemmomas and isolated neurofibromas are extremely rare. Here, a case of neurofibroma of the maxillary sinus. We present the case of a 60-year-old female patient with the chief complain of growth in the upper right back region of the jaw, which was preceded by exfoliation of teeth in the same region 1 month back.

Keywords: Maxillary sinus, neurofibroma, neurofibromatosis type 1

Introduction

Neurofibromas are benign tumors of peripheral nerves that arise from the connective tissue of their sheaths, especially the endoneurium that is composed of Schwann cells, perineural cells and fibroblasts. They are much more frequent in association with neurofibromatosis type 1, also known as von Recklinghausen's disease, although isolated cases may also appear.

Neurofibroma is considered to be one of the most common neurogenic tumor, but an uncommon intraoral neoplasm. Its most common site is the skin presenting as multiple fibromata as part of syndrome of neurofibromatosis (von Recklinghausen's disease) or polyglandular syndrome multiple endocrine neoplasia III.

Peak age of presentation is said to be third decade of life; however, it occurrence between 10 months and 70 years of age has been reported. Debilitrating or immunocompromising diseases are not known predisposing factors and sex predilection is contradictory.

Case Report

The present case report is about a 65-year-old female patient came to the out-patient department with the chief complaint of growth in the upper right back region of the jaw since 1 month.

Patient was apparently alright 1 month back than she noticed exfoliation of teeth in right upper back region of the jaw, which was followed by growth in the same region initially; the growth was small in size which gradually increased to present size of approximately 2.5 cm × 2.5 cm. A history of chewing tobacco 8-10 times a day since 45 years had been obtained from the patient. She presently experiences some difficulty in swallowing. Patient has no history of pus discharge, nasal congestion, nasal drainage or no feeling of heaviness on bending, also no pain or paresthesia associated with the growth.

On general examination, all vital signs were within the normal limits. The patient seemed to be well-oriented with time, place and person.

Extraorally, the face was asymmetrical due to diffuse swelling present on the right side of face extending anteroposteriorily from the right corner of mouth up to level of outer canthus of the right eye and superio-inferiorly from right ala of nose up to the base of the mandible having a size of approximately 2.5 cm × 2.5 cm, roughly oval in shape with a smooth surface.
having color similar to the adjacent area in the lesional site same as that of adjacent skin [Figure 1].

The swelling is firm to hard in consistency, non-tender, single submandibular lymph node of size 2 cm × 1 cm approximately firm in consistency, mobile, non-tender on palpation [Figure 2].

Intraorally, a single pedunculated growth is seen in the right alveolus in the maxillary posterior region extending up to the buccal vestibule mesio-distally, 1 cm from the midpalatine raphae up to 1 cm lateral to midline to the alveolus laterally and anteroposteriorly from distal of upper right first premolar up to upper right third molar region also, supero-inferiorly from the depth of vestibule up to 1 cm below the occlusal surface of maxillary teeth having a size of approximately 4 cm × 6 cm which is roughly oval in shape having bosselated surface with indentations that appear whitish cream in color with well-defined margins. On palpation, the swelling is soft to firm in consistency, non-tender along with grade 1 mobility with upper right first and second premolar.

On radiological examination
Orthopantomogram shows teeth present
5−1 | 1−8
8−1 | 1−7
Along with proximal caries with 67 along with moderate to severe interdental alveolar bone loss. Along with haziness in the right maxillary sinus region [Figure 3].

Peripheral nervous system view
There was the presence of slight haziness in the right maxillary sinus with increased separate in the sinus when observed in the PNS view.

On obtaining provisional diagnosis of fibroma a biopsy was conducted and soft-tissue excision was carried out by exposure of the right maxillary sinus by Caldwell-Luc approach from upper right first premolar to upper right third molar region along with curettage from right maxillary antrum.

The tissue was subjected to routine histological examination [Figure 4] and the hematoxylin and eosin stained slide showed histological features such as tumor composed of a myxoid stroma and spindle cellularity, with scant, poorly defined cytoplasm, with some areas of plexiform pattern and other more collagenized ones, with no signs of malignancy [Figure 5,6 and 7] Furthermore, for the identification of reticular fibers a Reticulin stain was done [Figure 8].

Discussion
Neurofibromas are benign tumors which are frequently associated with neurofibromatosis type 1, such as von Recklinghausen’s disease, although they can occur in isolation. The cell of origin for neurofibroma is believed to arise from the perineural fibroblasts which are neuroectodermal in origin. It is seen either as solitary form and multiple forms.}\end{quote}
They cause of solitary neurofibroma is unknown. Neurofibromatosis is inherited as an autosomal dominant trait with a high degree of penetrance but variable expressivity. In multiple neurofibromatosis, neurofibromas are encountered on both the skin and mucosal surfaces. Classified as nodular and diffuse form. Another classic feature of this disease is the presence of large diffuse macular brown pigmentation on the skin-café au lait spots.[2]

Neurofibroma is rare in the head and neck region when compared with schwannoma (25%). Solitary neurofibroma of maxillary sinus is exceedingly a rare tumor. This relative rarity is exemplified in a review by Robitaille et al. out of 15 cases in their series of peripheral nerve tumors involving the paranasal sinuses, four were neurofibromas,[3]

Neurofibromas have to differentiate from schwannoma and malignant peripheral nerve sheath tumors. Schwannoma shows distinctive areas identified as Antoni types A and B, typical palisade pattern of nuclei and tumor density is higher compared with the neurofibromas.[4] Nasal and paranasal neurofibromas arise from the ophthalmic and maxillary division of the trigeminal nerve. The symptoms are non-specific and depend greatly on the exact location and extent of the injury. They are, thus, often clinically silent, reaching considerable size in this location before diagnosis.[5]
Complete surgical excision remains the gold-standard treatment for neurofibromas. Total surgical excision in the head and neck region may pose a challenge to the surgeon due to infiltrative nature of the disease. Malignant peripheral nerve sheath tumors (2-5%) are characterized by hypercellularity and pleomorphic tumor cells and nuclei. [6]

Occasionally, such procedure may require sacrifice of vital structures leading to significant functional deficit or leave a cosmetic defect which increases the morbidity and mortality of the disease. [7] Although intra-lesional excision will reduce morbidity, it is usually associated with recurrence. [8]

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