

Neurofibroma of Maxillary Sinus: A Rare Case Report

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Abstract

Neurofibroma is a tumor of neuronal origin. Solitary neurofibroma of maxillary sinus is exceedingly rare tumor.

Keywords: Neurofibroma, maxillary sinus, malignant peripheral nerve sheath tumor (MPNST).

INTRODUCTION

Neurofibroma is a benign, slow growing, encapsulated, heterogeneous peripheral nerve sheath tumor that arises from the connective tissue of nerve sheaths, especially the endoneurium.¹ They may occur as isolated, sporadic lesions but are much more common in association with neurofibromatosis type 1, which is also known as Von Reckling Hausen's disease. It usually presents slow growing painless swelling. It is the histopathological study which usually reveals the true nature of the disease. Commonly involved nerves in the head and neck are the facial, the last four cranial nerves and the cervical sympathetic chain.² Neurofibroma is rare in head and neck region as compare to schwannoma (25%). Solitary neurofibroma of the maxillary sinus is exceedingly rare tumor.³ This relative rarity is exemplified in a review by Robitaille et al out of fifteen cases in their series of peripheral nerve tumors involving the paranasal sinuses, only four were neurofibromas.⁴ Rakshit et al in retrospective study of 266 cases of peripheral nerve tumor, found only one case of neurofibroma of nasal fossa.⁵ In the last twelve years, we have encountered one such case and is being reported here for clinical interest.

CASE REPORT

Twenty-five years male patient presented in ENT OPD with history of postnasal drip for the last 5 years off and on,

intermittent in nature, relieved with medical management and swelling of right cheek for the last 1 month. No history of nasal obstruction, epistaxis or headache. Examination of the nasal cavity showed spur on right side and mucopurulent discharge in the right nasal cavity. Middle turbinate was little boggy. No mass was seen. The examination of the facial region showed a bulge of the right cheek of approximately 3 cm in diameter with diffuse margin. It was firm in consistency and nontender. Clinically, a cyst of dental origin was suspected. X-ray of paranasal sinuses (Water's view) showed radiopaque shadow of right maxillary sinus (Fig. 1).



Fig. 1: Water's view



Fig. 2: Coronal view of CT scan of maxillary sinus

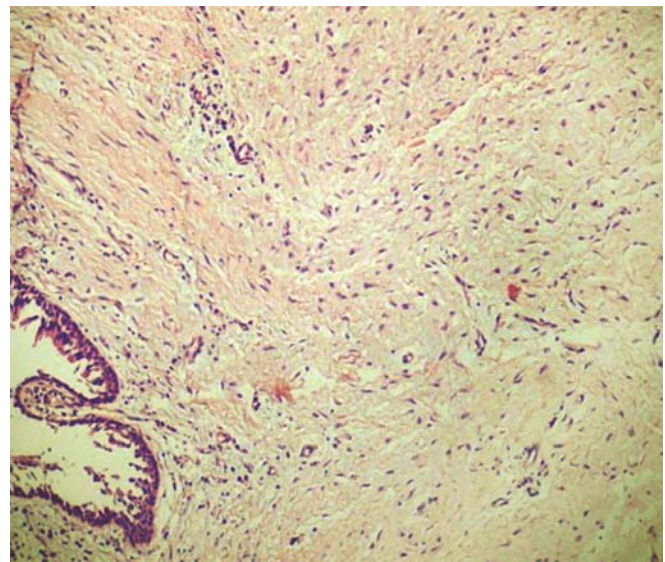


Fig. 4: Histopathological view (20X)

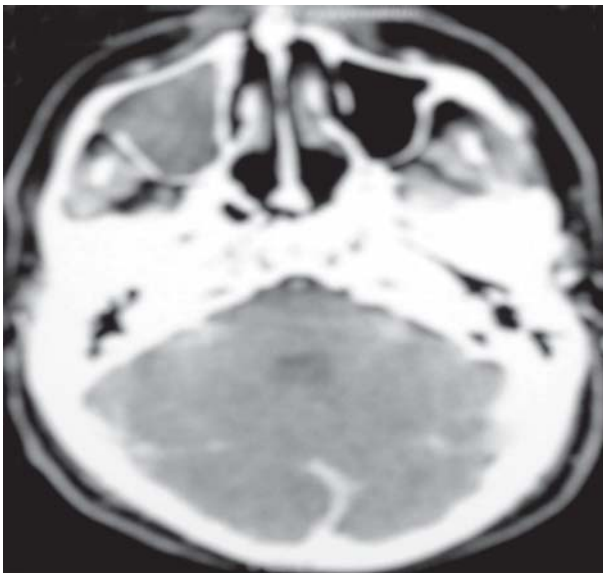


Fig. 3: Axial view of CT scan

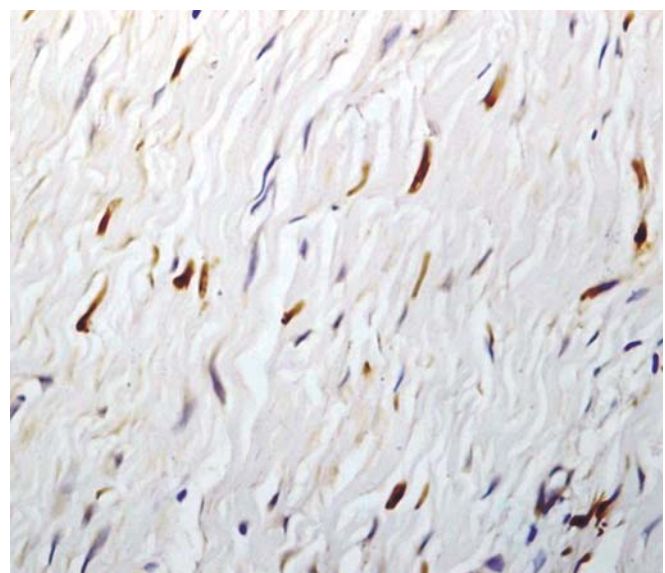


Fig. 5: Histopathological view (40X)

Computerized tomography of nose and paranasal sinuses showed heterogeneous soft tissue shadow in right maxillary sinus (Figs 2 and 3).

Exposure of the right maxillary sinus by Caldwell- Luc approach revealed blue colored thick wall cystic mass filling the maxillary sinus with attachment antero-inferiorly. The cystic mass was filled with caseous material. Mass removed in toto. Histopathological examination revealed neurofibroma (Figs 4 and 5).

There was no sign of recurrence after 6 months of follow-up.

DISCUSSION

The ophthalmic, maxillary division of trigeminal nerve and autonomic plexuses are the nerve of origin for neuro-

fibromatosis in the nose and paranasal sinuses.² Neurofibroma is divided into solitary and multiple that occur in association with neurofibromatosis-type 1. Neurofibromas may arise on any cranial nerve except the ophthalmic and optic nerve as they do not possess Schwann cell sheath.

Nerve sheath tumors of the maxillary sinus usually reach certain size before diagnosed clinically. Symptoms are nonspecific and depend heavily on the location and extension of the lesion.⁶ As in the presentation, patients with benign primary nerve sheath tumor of the trigeminal nerve often do not have any neurological deficits.

Preoperative CT scan show heterogenous soft tissue density and or destruction of paranasal sinuses.⁷ Micro-

scopically they are composed of Schwann cells, fibroblasts, perineural cells, axons and mast cells that are embedded in extended extracellular matrix.¹ Immunoreactivity for S 100 protein, neuron-specific enolase and vimentin is characteristic for neurofibromas.²

Neurofibromas have to be differentiated from schwannoma and malignant peripheral nerve sheath tumors. Schwannoma show distinctive areas identified as Antoni types A and B, typical palisade pattern of nuclei and tumor density is higher compared with the neurofibromas, which usually show a mucoid extracellular matrix with only scattered tumor cells.⁷ Malignant peripheral nerve sheath tumors are characterized by hypercellularity and pleomorphic tumor cells and nuclei, features not presented by this tumor.

Complete surgical excision is the therapy of choice.⁸ Follow-up is mandatory because the lesion may recur. Solitary neurofibromas very rarely undergo malignant change. The transformation of neurofibroma into malignant peripheral nerve sheath tumor has been observed in 2 to 5% of cases of neurofibromatosis type 1.¹

Despite the rareness of the disease, otolaryngologists should keep the possibility of a neurofibroma of the paranasal sinuses in mind.

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