

## A Rare Case of Diffuse Neurofibroma of Maxillary Sinus

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**Abstract:** Neurofibroma is a benign slow growing tumour of the peripheral nerve sheath; frequently associated with neurofibromatosis type I. Diffuse neurofibroma of maxillary sinus is an exceedingly rare tumour.

We report a case of a 17-year-old male patient presented in the ENT department with a painless swelling of the left side of the face for the last one year. CT scan revealed a soft tissue tumour with involvement of predominantly left premaxillary region. It also showed bilateral maxillary, right ethmoidal and sphenoid sinusitis with DNS and bilateral inferior turbinate hypertrophy. FNAC of the swelling showed cytomorphological features suggestive of benign mesenchymal lesion possibly benign nerve sheath tumour. Tumour excision was done. On histopathological examination, it consisted of elongating spindle cells with poorly defined, pale eosinophilic cytoplasm and tapering wavy nuclei in a fibromyxoid matrix. Many foci of meissnerian differentiation were identified. Dendritic cells containing melanin pigments were also found. The features were consistent with diffuse neurofibroma. This case proves how aggressive the tumor of this type may be, showing few clinical signs. Despite the rareness of this disease, pathologists should keep this diagnosis in mind within the range of tumours of the paranasal sinuses.

**Keywords:** maxillary sinus, von Recklinghausen's disease, neurofibroma

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### I. Introduction

Neurofibroma is a benign tumor originating from peripheral nerve sheath. In contrast to schwannoma, neurofibroma appears to originate within the endoneurium, and, in fact, a small subset of neurofibromas are entirely intraneural.(1). 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.. It is most commonly present in the skin 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. Histologically, neurofibroma has been broadly classified into: 1. Solitary 2.diffuse 3. Plexiform .

Intramaxillary neurofibroma is an extremely rare occurrence. Intramaxillary neurofibromas may present as sessile or pedunculated, progressive but slow growing lesion. Often painless but may be painful due to pressure effects.[2,3,4] .Here we report a rare case of solitary neurofibroma of the left maxillary sinus in a 17 year old patient and try to describe the clinical, histopathological and radiological characteristics of this tumour.

### II. Case Report

A 17 year old male patient presented in the ENT department with a painless swelling of the left side of the face for the last 1 year.

On general examination, all vital signs were within normal limit. Externally, the face was asymmetrical due to diffuse swelling present on the left side of the face extending anteroposteriorly from the left corner of the mouth up to the level of outer canthus of left eye and superior inferiorly from right ala of nose upto the mandible measuring ( 4X4) cm (fig 1) . The growth is roughly oval in shape with firm to hard in consistency.

Endobuccal examination revealed a tumefaction of the left upper gum with palatal and vestibular extension.

Ophthalmic examination showed posteriorly displaced left eyeball with painful ocular mobility on down and outward gazes. The visual acuity of the both eye was normal. There was corneal dryness in the inferior half of left eye.

Endoscopic examination of the nose revealed DNS with convexity towards right side and bilateral inferior turbinate hypertrophy.

CT scan revealed a soft tissue tumour measuring about 8.2cm x4 cm, noted in left premaxillary region showing minimal enhancement on contrast study. The lesion is insinuating between the posterior margin of left maxillary sinus and left temporalis sinus, extending medially upto bridge of the nose, superiorly abutting roof of nose, inferiorly upto the angle of left mandible and posteriorly extending upto the anterior wall of left maxillary sinus. There is also evidence of bilateral maxillary (right > left), left ethmoidal and sphenoidal sinusitis (fig 2, 3).

FNAC of the swelling showed scattered plump fibroblast, few clusters of acinar gland. There were few spindle cells with pointed nuclei adhered with pinkish fibrillary material .The above cytomorphological features suggested of benign mesenchymal lesion possibly benign nerve sheath tumour.

Multiple biopsy of the swelling showed circumscribed but unencapsulated lesion replacing dermis and subcutis consisting of elongating spindle cells with poorly defined, pale eosinophilic cytoplasm and tapering wavy or buckled nuclei, admixed with indeterminate short spindle cells (fig4, 5). The mixed components were in a fibromyxoid matrix. Many foci of meissnerian differentiation were identified. Dendritic cells containing melanin pigments were found (fig 6). Diffuse manner in which the lesion infiltrates subcutaneous fat superficially was also seen (fig 7). The features were consistent with diffuse neurofibroma.

On the basis of the above diagnosis, tumour excision was done..

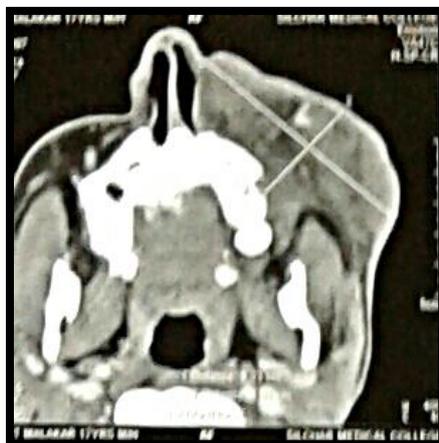
Gross examination of the excised specimen showed a firm greyish mass of size (5X4X4) cm<sup>3</sup>. There was no capsule adherent to the mass. Numerous nodules protruding from the outer surface of the lesion was seen (fig 8)

Final excisional histopathological examination also showed features of diffuse neurofibroma

#### 1.1 PICTURES:



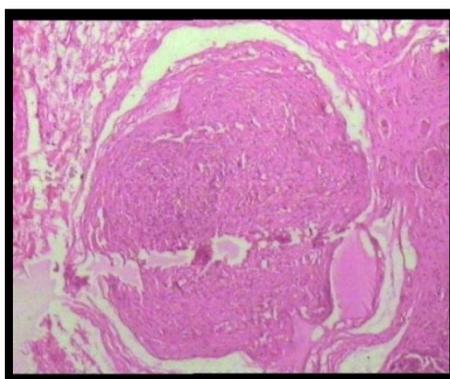
**figure 1:** diffuse swelling present on the left side of the face



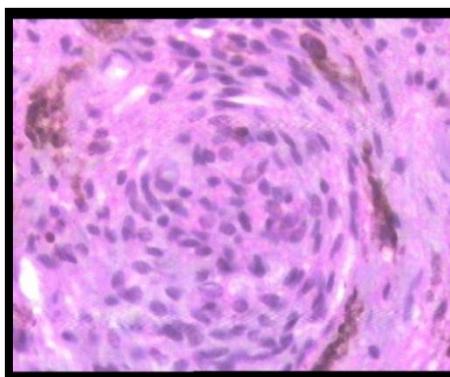
**figure 2:** ct scan of faciomaxillary region axial contrast view



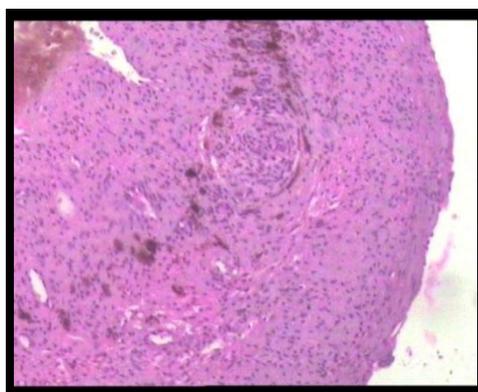
**figure 3:** ct scan of faciomaxillary region axial non contrast view



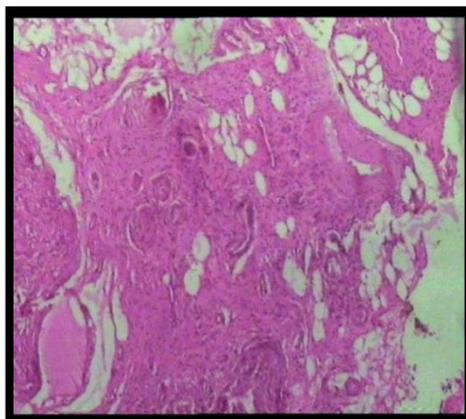
**figure 4:** microscopic low power view showing diffuse replacement of the dermis and subcutis by neurofibromatous tissue



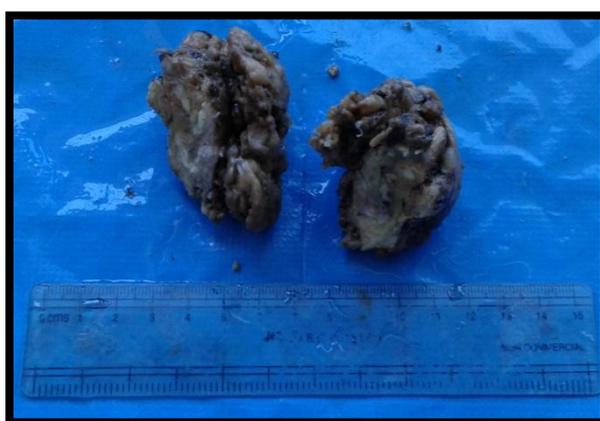
**figure 5:** microscopic high power view showing elongated spindle cells with poorly defined, pale eosinophilic cytoplasm and tapering.



**figure 6:** microscopic lower power view showing dendritic cells containing melanin pigment



**figure 7:** microscopic low power view showing diffuse manner in which these lesions infiltrate subcutaneous fat



**figure 8:** gross examination of specimen shows greyish mass of size ( 5x4x4 )cm<sup>3</sup>

### III. Discussion

Neurofibroma is a benign nerve sheath tumour which arises from the peripheral nervous system. It is broadly divided into solitary and multiple. The last one occurs more frequently especially in association with neurofibromatosis type 1, such as Von Recklinghausen's disease. When compared with schwannoma (25%), they tend to be more common. Neurofibroma is rare in the head and neck region. Diffuse neurofibroma of maxillary sinus is an exceedingly rare tumor. This relative rarity is exemplified in a review by Robitaille *et al.* in which out of 15 cases of peripheral nerve tumors involving the paranasal sinuses in their series, only four were neurofibromas. Neurofibromas may arise on any cranial nerve except the optic nerve as they do not possess Schwann cell sheath. Nasal and paranasal neurofibromas arise from the ophthalmic and maxillary division of the trigeminal nerve [5]. The symptoms are non-specific and depend greatly on the exact location and extent of the lesion [6]. In this location, it is often clinically silent until reaching considerable size before diagnosis. No neurological deficit is present in trigeminal nerve tumour. However, bone destruction is a common feature of these lesions. CT scan show heterogeneous soft tissue density with destruction of the paranasal sinuses.

Microscopically, diffuse neurofibroma is characterized by diffuse replacement of the dermis and subcutis by neurofibromatous tissue with consists of elongated spindle cells with poorly defined, eosinophilic cytoplasm and tapering, wavy or buckled nuclei, admixed with more indeterminate short spindle cells, numerous small nerve fibers, and frequent mast cells. These mixed components are set in a variably fibromyxoid matrix. In virtually all cases, foci of meissnerian differentiation are identified. The constituent nerves (or nerve fibers) tend to appear markedly hypertrophic and edematous. It is common to find dendritic cells containing melanin pigment in these tumors. Occasionally multinucleate giant cells of uncertain origin are also encountered. The diffuse manner in which these lesions infiltrate subcutaneous fat superficially may be mistaken for that of a dermatofibrosarcoma. Most of the time, neurofibroma has to be differentiated from schwannoma.

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. Occasionally, such procedure may require sacrifice of vital structures leading to significant functional

deficit or leaving 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]

#### **IV. Conclusion**

This case proves how aggressive the tumor of this type may be, while showing few clinical signs. At the beginning symptoms are uncharacteristic, such as sporadic neuralgia or periodical parestheses of the tissues connected to the particular nerve. However, symptoms become stronger with the tumor's growth and damage done to nearby structures which depends on the tumor location. It is also important to pay attention to the single tumor as it may be the beginning of type 1 neurofibromatosis. Despite the rareness and the difficulty diagnosing this disease, otolaryngologists and pathologists should keep this diagnosis in mind within the range of tumours of the paranasal sinuses.

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