NEUROFIBROMA OF PARANASAL SINUSES - A CASE REPORT

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ABSTRACT : *Primary neurogenic tumors in nose and paranasal sinuses are unusual entities. The majority of the reported cases have dealt with neurilemmomas and isolated neurofibromas are extremely rare. Here a case of neurofibroma of the paranasal sinuses is described.*

Key Words : *Neurofibroma, Paranasal Sinuses*

INTRODUCTION

Neurofibromas of PNS are a rare entity. The ratiry of this disorder enhances the diagnostic difficulty preoperatively and histological evaluation later.

Schwann cell tumors are classified as neurilemmoma, neurifibroma and malignant schwannoma. Though those neurogenic tumors are found in the head, neck and flexoral surfaces of the upper and lower limbs, their affection of paranasal sinuses in the reported literature have been far and few[1,2].

In this paper a case of neurofibroma of the paranasal areas is described due to its rarity.

CASE HISTORY

A 38 years old male came to us with a cheek swelling on the right for 5-6 years duration. The swelling had insiciously increased in size and there was no history of trauma, teeth extraction or epistaxis, apart from a dull aching sensation for last 5-6 months and right sided partial nasal obstruction for 1 year.

On examination, as the swelling was present over the right cheek which was firm measuring 7.5 cm x 7.5 cm., the nasofacial furrow (Rt) was obliterated but the infraorbital margin was not dehiscent (Fig. I). On the inner aspect, the gingivolabial sulcus was obliterated and the tumor growth extended from right 2nd molar to the other side of central incisor. There was no palatal bulge. ANS revealed a medial shift of the lateral wall of the right nasal cavity producing a marked narrowing of the right nasal cavity. There was no anaesthsia over the cheek or inside the palate.

After admission, an incisional biopsy was performed but no definitive diagnosis could be made from the tissue.

The haematological examination and chest radiograph were within normal parameters.

CT scan showed a mass in Right maxillary antrum with bony erosion and anterior extra-antral soft tissue extension (Fig. II). The extension of the mass was also seen in right cheek and nodular infiltration was seen across maxillary hard palate extending into contra-lateral antral cavity (Fig. III**).**

The mass was explored by a sublabial approach. The mucosa along with remaining periosteal layer was elevated and a mass was seen eroding the alveolar process of both maxilla and nasal spine, anterior part of maxillary crest of septum and part of medial and anterior wall of right maxilla. The exterior portion had a myxomatous appearance but deeper debulking revealed much more friable tissue. It also extended into the anterior and posterior group of

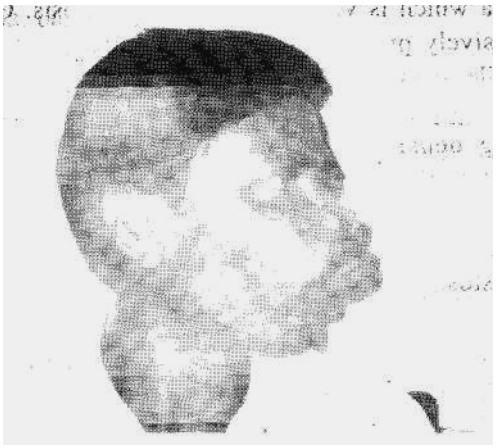

Fig. I : Clinical photograph showing right sided cheek swelling with obliteration of naso-facial furrow.

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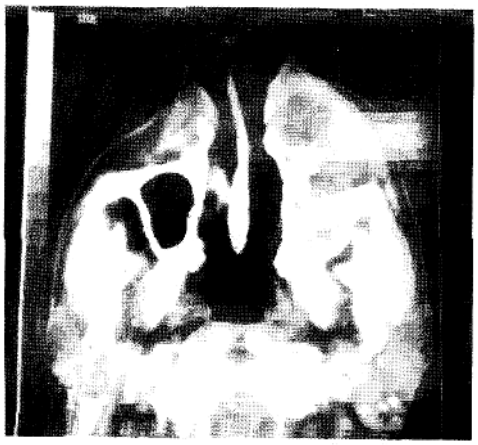

Fig. II : Photograph showing CT scan of the patient with tumor in right antrum extending sublabially by breaching anterolateral wall (Axial view).

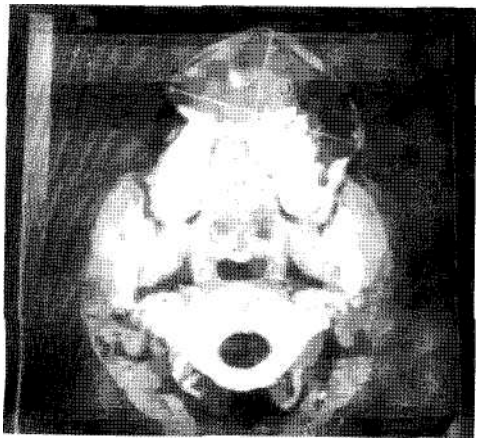

Fig. III : Photograph of CT scan showing sublabial extension of the growth to opposite antrum (Axial view).

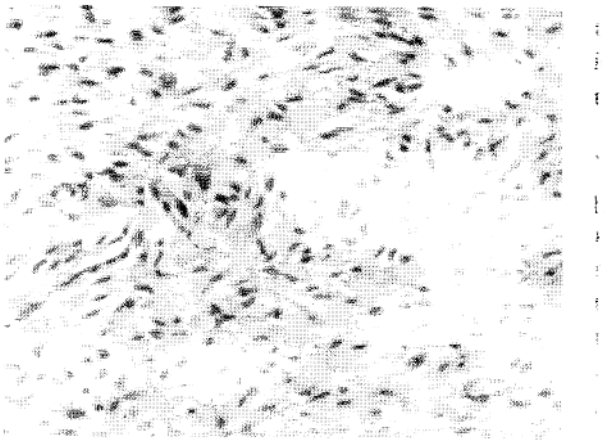

Fig. IV : Microphotograph shows proliferation of spindle cells with sprinkling of lymphocytes lying on a loose stroma. (H & E x 235)

Fig. V : Clinical photograph of the patient during the 6th post operative month.

ethmoidal air cells which were removed by endoscope. The operative specimen had a neoplastic appearance but the bleeding was not disproportionate. The posterior ethmoidal cells were not very enthusiastically curetted to prevent any injury to the optic nerve. An inferior antrostomy was performed before closing the wound after packing the antrum.

Postoperatively, the histopathology report of the operated specimen came as follows :-

Section showed stellate cells and spindle cells arranged in fascicular pattern lying on a loose oedematous myxoid background. Degenerated cells were also present. Bundle of wavy nucleoli with palisading patterns were seen. Hence the histopathological examination from this was considered to be neurofibroma. The patient has turned up twice in our clinic for follow up and no recurrence has been seen for last 8 months (Fig. V).

DISCUSSION

Schwann cell tumors occur infrequently in the nasal cavity and paranasal sinuses. This is surprising considering the fact that schwann cell neoplasms are found with moderate frequency elsewhere in the body. While judging from existing literature, they are unusual entities in these areas.[2]

A review of American and European literature till 1975 revealed 12 neurilemma 4 neurofibromas and one probable malignant schwanoma. A review of 4300 pathology specimen by Perzin et al revealed 6 neurofibromas. 4 malignant schwannomas and 2 nrurilemmomas involving nasal cavity, paranasal sinuses and nasopharynx. In a review of 152 benign and malignant neurilemmomas of head and neck, Kragh et al described 5 in the nasal fossa and antral

region. Thus only a small data of Schwann cell neoplasms involving the upper respiratory passage have been recorded.

The tumors are usually solitary and among the site of predilection in these areas combined nasal ethmoid complex is most frequent followed in order by maxillary sinus, intranasal and sphenoid sinus. In nearly all examples, the tumors have been solitary. Only in 10% of cases, the tumors are in association with Von Recklinghansen's disease (Geschitckter, 1935).[1] It is to be noted while neurilemmoma is benign, malignant transformation may occur as in neurofibroma[1,2,5] but exact incidence is difficult to determine and has been estimated at 12% of solitary[1] and in approximately 8% patients with Von Recklinghausens disease.[4]

The affected age group is from 25 to 55 years of life. The symptomatology is usually non-specific like epistaxis (more in nasal fossa and ethomoidal sinus), nasal obstruction, facial pain (especially maxillary tumors), haemorrhagic necrosis and thrombi are also responsible for misdiagnosing them as vascular tumors like angiofibroma or fibrotic nasal polyps. Thus, when not in association with VonRecklighausen's disease, they are difficult to diagnose clinically,

A few pertinent features regarding neurofibroma must also be kept in mind, that in very large tumor, excision may be mandatory and the nerve of origin may not be detailed. Secondly, since both benign and malignant change of nerve cell tumors erodes bone, it may not indicate malignancy per se and intracranial extension can complicate the natural course of both varieties of tumors. But since cosmetic and functional aspect of face should be kept in mind radical surgery may be postponed because even if the surgery is incomplete, recurrence may be after several years and hence this excessive curettage was not done in the post ethmoidal region.

The patient was reviewed for two follow ups at 3 and 8 months interval and he has been symptom-free and did not demonstrate any sign of recurrence.

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