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NEUROFIBROMA OF NASAL VESTIBULE: A RARE CASE REPORT

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ABSTRACT: Head & neck region is the most common site of neurogenic tumors like neurofibroma. Mostly they are multiple & associated with NF-2 syndrome. Solitary neurofibroma involving nose & paranasal sinuses are very rare. Here, we present a rare case report of solitary neurofibroma present over nasal vestibule of an 18 years aged old patient. If neurofibroma is present as a small solitary tumour, it is curable by adequate primary excision (Batsakis 1979). We discuss the clinical presentation, differential diagnosis, imaging characteristics and treatment of this rarely encountered lesion. This is important to consider such a rare variety of tumor in differential diagnosis of a unilateral soft tissue mass of nasal cavity.

KEY WORDS: Neurofibroma, Vestibule, Endoscopic endonasal surgery

INTRODUCTION: Neurofibroma is a benign neurogenic tumour arising from Schwann cells or peripheral tissues of nerve sheaths. It is usually presented with Von Recklinghausen's disease rather than as a solitary tumour¹. About 25% neurofibromas involve head & neck region and only 4% involve nose & paranasal sinuses². The potential for malignant transformation is 2.6% & in syndrome it varies between 3 & 15%³.

The treatment of choice for neurofibroma in the nose and paranasal sinuses is complete excision via a lateral rhinotomy approach or a mid-facial degloving approach⁴. However, if neurofibroma is present as a small solitary tumour, it is curable by adequate primary excision¹. Recently, the technique of endoscopic nasal surgery has rapidly developed, and transnasal endoscopic excision of the primary lesion is successful when neurofibroma is a solitary tumor.

Here we present a case report of solitary neurofibroma involving nasal vestibule of an 18 years patient.

CASE REPORT: The patient aged about 18 years came to our hospital with the chief complaint of progressive left side nasal obstruction, tingling sensation over nasal vestibule and bridge of nose of one year duration. He had no rhinorrhoea, epistaxis, anosmia, facial pain or headache. There was no history of recent nasal trauma. His medical and family histories were unremarkable. He was of average body built with no systemic complaints.

Anterior rhinoscopy, revealed a large sessile mass completely filling the left nasal cavity (Figure 1). The mass was firm in consistency, non tender, smooth surfaced and appeared to be covered by normal nasal mucosa. It did not bleed on touch.

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Figure 1

Figure 2

Figure 3

Diagnostic endoscopy revealed a firm, non lobulated, non tender, non bleeding mass filling the left nasal cavity upto superior part of inferior turbinate. The mass had a polypoidal mucosal surface, was ovoid in shape, and had a pale colour. It originated from left nasal vestibule. There was no posterior extension of the mass & the right nostril was normal. Nasopharynx was normal.

Plain paranasal sinus X-rays revealed no abnormality. CECT of nose & paranasal sinus was done to know the exact size and extension of the mass. CT shows slightly contrast enhanced soft tissue mass arising from left nasal vestibule (figure 2). There was no bony erosion or involvement of any important nearby structure.

On the grounds of the pre-operative CT scan and diagnostic endoscopic findings, the tumor seemed to be of benign nature. The mass was removed by transnasal endoscopic surgery for definitive diagnosis and treatment. The origin of mass was from the left nasal vestibule. Exision done along with a part of adjacent normal tissue. There was no much bleeding intraoperatively. Post-excisional mass was friable, firm of size 2x3cm (figure 3). On cut section it was pale, gray-white, homogenous & of firm consistency. Histological examination of the mass showed combined proliferation of eosinophilic, thin, wavy collagen fibers lying in a disorderly pattern and ovoid to spindle-shaped cells with a wavy configuration. The stroma was infiltrated with lymphocytes and mast cells without any mitotic activity.

The final pathological diagnosis was neurofibroma arising from nasal vestibule. The post-operative course was uneventful. With one year follow up there was no recurrence of mass.

DISCUSSION: Peripheral nerve-sheath tumours are divided into neurofibroma, schwannoma and neurogenic sarcoma. Neurofibroma and schwannoma are classified as benign, and neurogenic sarcoma as malignant⁵. All peripheral nerve-sheath tumours are believed to arise from Schwann cells. Neurofibromas are most commonly found in head and neck areas & flexoral aspects of extremities. However, they are rarely found in the nasal cavity and paranasal sinuses. The neurofibroma occurring in the nose may be solitary or multiple. Solitary neurofibroma arises along a nerve trunk with a spontaneous onset. In the nose and paranasal sinuses, the tumour arises from the first and second division of the trigeminal nerve and from autonomic plexuses, but cannot arise from the olfactory nerve, which has no Schwann cells¹. The definitive neural source arising from nasal vestibule is difficult to establish. There is no sex or racial predilection. A review of American and European literature till 1975 revealed twelve cases of neurilemoma, four cases of neurofibromas and one of probable malignant schwannoma. A review of 4300 pathology specimens by perzin et al revealed 6 neurofibromas, 4 malignant schwannomas and 2 neurilemmomas involving nasal cavity, paranasal sinuses and nasopharynx⁶. In a review of 152 benign and malignant neurilemmomas of

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head and neck, Kragh described five in the nasal fossa and antral region⁷. There is no sex or racial predilection⁹. Clinically, symptoms and signs are dependent on the site, extent and nerve of origin. Usually common symptoms and signs are nasal obstruction, epistaxis, facial pain, swelling, proptosis, rhinorrhea, anosmia & serous otitis media. Due to its indolent growth pattern, when it becomes huge in size, it may destroy bony barrier and can cause intracranial complication. Schwannomas often produce a painful sensation and tenderness and are predominantly centrifugally distributed, whereas neurofibromas are asymptomatic and have a primarily centripetally location. The diagnosis of neurofibroma is usually made only when the lesion is biopsied, because of the non-specific nature of these symptoms and signs.

It is important to distinguish between neurofibroma & schwannoma as neurofibroma is prone for malignant transformation⁸. Histologically both have almost similar feature, however schwannoma is encapsulated and has palisade arrangement. The neurofibroma is non-encapsulated, poorly circumscribed with an ill-defined margin. This is typified by relatively hypocellular proliferation of bland, pale eosinophilic spindle cells with wavy nuclei in a myxoid background. Usually, mitoses are not found. It is also important to distinguish it from malignant peripheral nerve sheath tumor, other spindle cell tumor like fibrosarcoma, synovial cell sarcoma and osteosarcoma.

Surgical resection with a preservation of normal anatomical landmarks is the sole treatment of choice of solitary neurofibroma presenting in the nasal cavity. Endoscopic endonasal surgery is the treatment of choice. When the lesion is inaccessible, lateral rhinotomy may be considered. After surgery it is necessary to follow up the case due to the chance of malignant transformation & recurrence.

CONCLUSION: Peripheral nerve sheath tumor like neurofibroma should be kept in mind while evaluating a case of soft tissue tumor of nasal cavity. Complete excision is the treatment of choice, however a close follow up is essential to rule out any recurrence of the tumor. Although their occurrence does not seem to be very high in number, still a vigilant attitude on the part of the surgeon keeping in mind it as a differential diagnosis will help us to identify such tumors more confidently and accurately.

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