International Journal of Otorhinolaryngology and Head and Neck Surgery
Malhotra V et al. Int J Otorhinolaryngol Head Neck Surg. 2024 Dec;10(6):746-748
http://www.ijorl.com
pISSN 2454-5929 | eISSN 2454-5937

DOI: https://dx.doi.org/10.18203/issn.2454-5929.ijohns20243520

Case Report

An incongruous occurrence: rare case of a huge sinonasal neurofibroma

Vikas Malhotra[1], Deepika Sethi[1], Nidhi Mahajan[2], Moudipa Chatterjee[1], Nitish Aggarwal[1]*

[1]Department of ENT, Dr. Baba Saheb Ambedkar Medical college and Hospital, Rohini, Delhi, India
[2]Department of Pathology, Dr. Baba Saheb Ambedkar Medical college and Hospital, Rohini, Delhi, India

Received: 25 May 2024
Revised: 18 October 2024
Accepted: 04 September 2024

***Correspondence:**
Dr. Nitish Aggarwal,
E-mail: nitishaggarwal28@gmail.com

ABSTRACT

Neurofibromas (NF) are benign peripheral nerve sheath tumors that arise from schwann cells and intraneural fibroblasts. Solitary NF are extremely rare with only a few reported cases in the literature that involve the nasal cavity and paranasal sinuses. Due to its slow growth and the non-specific nature of the symptoms, its clinical presentation and diagnosis is often delayed. Here we present a case of a 65-year-old female with left sinonasal neurofibroma which was surgically managed in our department successfully. It was 9.5 cm in greatest dimension and weighed 380 gm which is the largest amongst the available published reports that we could find.

Keywords: Sinonasal neurofibroma, Nasal rumours, Lateral rhinotomy, Nasal masses

INTRODUCTION

Neurofibromas (NF) are benign peripheral nerve sheath tumors that arise from schwann cells and intraneural fibroblasts.[1] It can occur either as multiple lesions in individuals affected by neurofibromatosis type 1 (NF1) or as a solitary neurofibroma. Solitary NF without associated von Recklinghausen's disease are extremely rare with only a few reported cases in the literature. 25% to 45% of NF arise in the head and neck region with only 4% involving nasal cavity and paranasal sinuses.[2] Due to its slow growth and the non-specific nature of the symptoms, its clinical presentation and diagnosis is often delayed.[3] The present case reports a prodigious sinonasal neurofibroma treated successfully in the department of ENT.

CASE REPORT

A 65-year-old home maker presented to ENT outpatient department with a history of severe nasal obstruction more on the left side as compared to right. It was first noticed around four years back and was clinically diagnosed as a nasal polyp. Further evaluation and surgical management was planned for the same. However, due to COVID pandemic the patient decided to not go ahead with it. Patient also complained of nasal discharge and occasional hemicranial headache, which was relieved on taking analgesics. No significant family history was revealed.

Anterior rhinoscopy showed bilateral extensive synechiae. However, a previous diagnosis of nasal polyposis urged us to plan a diagnostic nasal endoscopy with biopsy. Contrast enhanced CT of the PNS showed a heterogenous soft tissue lesion of 9.3×3.8×3.6 cm (APXTRXCC) filling up the right nasal cavity, nasopharynx and choana with partial erosion of middle and inferior turbinates with obstruction of sphenoethmoidal complex and the osteomeatal complex (Figure 1). The patient was evaluated for surgical fitness and the mass was excised via a lateral rhinotomy approach due to extensive synechiae limiting the operating field in endoscopy (Figure 2). The mass was excised in toto and seemed to arise from the nasal cavity per se and not form the paranasal sinuses. The patient tolerated the procedure well and postoperative period was uneventful. Grossly, tumor was multinodular with

glistening surface and on cut section, firm with no haemorrhage or necrosis (Figure 3). Histopathological examination revealed a well circumscribed tumor composed of spindled out cells arranged in fascicles and sheets with curvilinear and wavy nuclei (Figure 3). These cells were dispersed in a collagenized stroma and few mast cells. No nuclear palisading, nuclear pleomorphism, mitosis or necrosis was identified. Tumor cells showed strong immunohistochemical expression of S-100 and negative for SMA, CD34, STAT 6, CK and HMB45. A final diagnosis of sinonasal neurofibroma was made.

The patient was then thoroughly examined clinically but did not reveal any other swellings in the body. The patient was on regular follow up and no local recurrence was seen. Extensive literature search revealed this to be the largest neurofibroma excised so far.

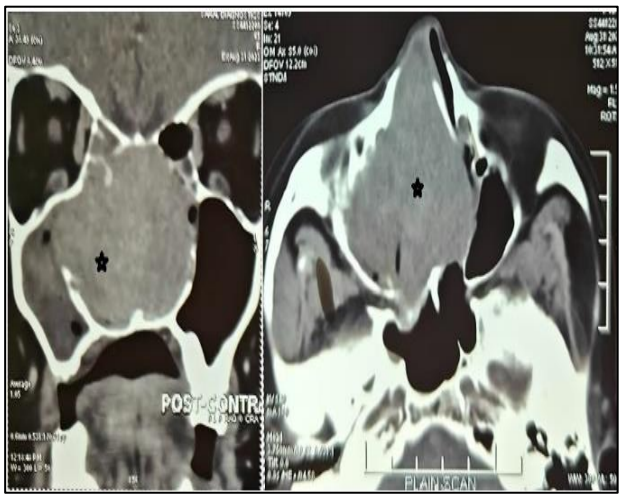

Figure 1: CECT of the PNS of a heterogenous soft tissue lesion filing up the right nasal cavity, nasopharynx and choana.

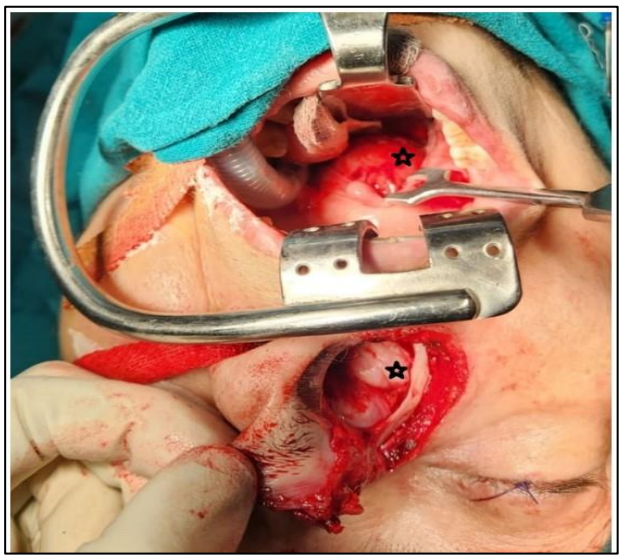

Figure 2: Intraoperative image of the mass being excised via a lateral rhinotomy approach.

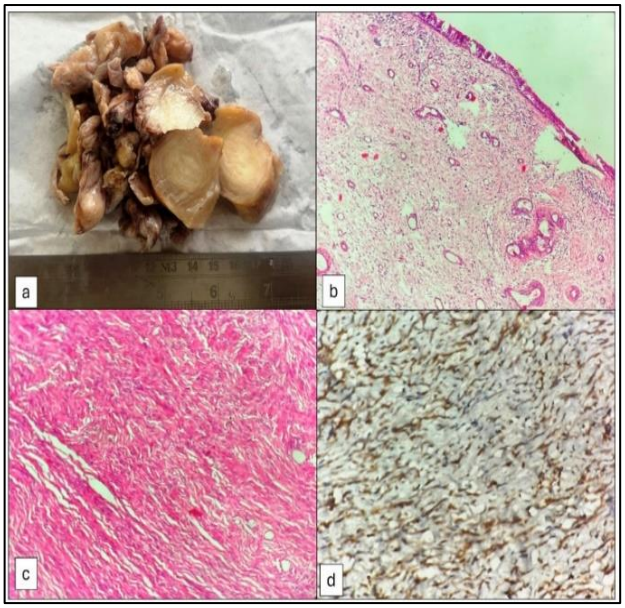

Figure 3 (a-d): Macroscopic image of the tumor composed glistening grey white soft tissue pieces. Focal area of hemorrage seen. Microscopic image tissue lined by respiratory epithelium and underlining unencapsulated lesion composed of spindled out cells (H and E, 40×). High power view of spindled out cells with fibrillary cytoplasm. No nuclear pleomorphism or mitosis (H and E, 100×). Immunohishochemistry shows tumor cells expressing S-100 (DAB-chromogen, 100×).

DISCUSSION

Benign peripheral nerve sheath tumours include neurilemomas (schwannomas), malignant schwannomas and less frequently, NF. They are broadly classified into dermal and plexiform types. Dermal type are associated with individual peripheral nerves and plexiform types are usually associated with many nerve bundles.[4]

The chances of finding these tumours in the sinonasal tract is exceptionally rare with only a few reported cases in the literature. NF are most common in nasal cavity (64%), followed by the maxillary sinus (17%), or mixed sites.[5]

A series of these cases by Azani et al found the mean age to be 46.2 years (ranging from 26 to 75 years).[5] The patient in our case was a 65 year old female. They found the tumour to be slow growing with a mean duration of symptoms of 42.9 months, quite similar to current case.

Azani et al in their series found the tumor size to range from 0.4 to 4.1 cm with a mean of 2.2 cm.[5] Current case was 9.5 cm in the greatest dimension and weighed 380 grams which is the largest amongst the available published reports that we could find. Histopathological examination typically showed features characteristic of neurofibroma.

A thorough diagnostic nasal endoscopy followed by radiological evaluation is a must for all cases of suspected nasal tumours as they are frequently misdiagnosed as nasal polyposis in less experienced centres.

Surgical excision forms the mainstay of treatment. The type of operation employed is dependent on the extent and location of tumor. Complete local excision is usually curative, and prognosis is excellent, if tumor is completely removed.[6] Endoscopic approach, nowadays is significantly more common However, external approaches (e.g., midfacial degloving, lateral rhinotomies) are excellent techniques for larger tumours and cases with abnormal nasal cavity as seen in our case where patient was successfully treated using a lateral rhinotomy approach.

Histologically, the neurofibroma is unencapsulated, poorly circumscribed with an ill-defined margin. It is characterized by relatively hypocellular proliferation of bland, palely eosinophilic spindle cells with wavy nuclei in a copious fibrillary or rather myxoid background. It shows immunoreactivity of S-100 protein, neuron specific enolase (NSE), and vimentin but not for Desmin, or smooth muscle actin and are useful in differentiating from other mesenchymal tumors.[1,2,5] Similar histological findings were observed in our case as well.

The differential diagnosis includes lymphoma, inverted papilloma, intranasal extension of juvenile angiofibroma, schwannoma, septal dermoid, idiopathic midline granuloma, and malignant tumors.[7]

If not excised completely, NF may recur locally and require further local resection. Malignant transformation of neurofibroma is rare unless the patient has NF1 or 2. Our patient was on regular follow up and did not show any signs of recurrence on repeated endoscopic evaluations.

CONCLUSION

Sinonasal NF are rare tumours which are often missed due to its slow growth. Surgical excision forms the mainstay of treatment with lateral rhinotomy approach being a good surgical option.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Prakash MD, Viswanatha B, D'Souza GE. A rare case of isolated nasal neurofibroma mimicking sinonasal polyp. Res Otolaryngol. 2014;3(3):49-51.
2. Hillstrom RP, Zarbo RJ, Jacobs JR. Nerve sheath tumors of the paranasal sinuses: electron microscopy and histopathologic diagnosis. Otolaryngol Head Neck Surg. 1990;102(3):257-63.6.
3. Perzin K, Panyu H, Wechter S. Nonepithelial tumors of the nasal cavity, paranasal sinuses and nasopharynx: A clinicopathological study. XII: Schwann cell tumors (neurilemoma, neurofibroma, malignant schwannoma). Cancer. 1982;50:2193-202.
4. Adnane C, Adouly T, Elhani L, Mahtar M. Solitary neurofibroma of the maxillary sinus: A rare case report. J Head Neck Physicians Surgeons. 2014;2(2):136-41.
5. Sharma SB, Hong P. Solitary neurofibroma of the frontal sinus. Case Rep Otolaryngol. 2012;2012.
6. Azani AB, Bishop JA, Thompson LD. Sinonasal tract neurofibroma: a clinicopathologic series of 12 cases with a review of the literature. Head Neck Pathol. 2015;9:323-33.
7. Riveros AC, Proietti V, Serrano C, Ruggeri CS. Sinonasal Schwannomas. J Otolaryngol Rhinol. 2021;7:108.
8. Yap D, Fox H, Addams-Williams J. Unusual case of a Proptosed eye: isolated right Maxillary Neurofibroma. Case Rep Pathol. 2016;2016:4294729.

Cite this article as: Malhotra V, Sethi D, Mahajan N, Chatterjee M, Aggarwal N. An incongruous occurrence: rare case of a huge sinonasal neurofibroma. Int J Otorhinolaryngol Head Neck Surg 2024;10:746-8.