

A Rare Case of Isolated Nasal Neurofibroma Mimicking Sinonasal Polyp

Mylanahalli Doddarangiah Prakash, Borlingegowda Viswanatha*, Glen Edwin D'Souza

Otorhinolaryngology department, Bangalore medical college & research institute, Bangalore, India

Abstract Neurofibromas are benign peripheral nerve sheath tumors which originate from Schwann cells. They are known to occur in extremities as a local manifestation of Von Recklinghausen's disease i.e. type 1 neurofibromatosis, but isolated neurofibroma occurring in the nasal cavity is a rare entity. Here we report one such case in a 60 year old lady who, presented to us with complaints of difficulty in breathing and a mass protruding from her right nasal cavity. Clinical diagnosis of inverted papilloma was made. Computed tomography suggested a polyp like mass lesion in the right nasal cavity, which was excised in toto by endoscopic endonasal approach. The mass was histologically confirmed to be a neurofibroma. Isolated neurofibroma, even though a rare entity in the nasal cavity should be thought of when history and clinical examination is not classical of other well-known nasal masses like sinonasal polyps or papillomas.

Keywords Nasal neurofibroma, Sinonasal polyp

1. Introduction

Neurofibromas are benign peripheral nerve sheath tumors. They arise from schwann cells and fibroblasts of the connective tissue of the endothelium. They are broadly classified into dermal and plexiform types. Dermal type is associated with individual peripheral nerves and plexiform types are usually associated with many nerve bundles [1]. Of the 25% to 45% neurofibromas that arise in the head and neck region, only 4% involve the nasal cavity and paranasalsinuses [2]. Solitary neurofibromas without associated von Recklinghausen's disease are rare. They have a malignant potential of 2.6% and if associated with von Recklinghausen's disease, the chances for malignant transformation are 3-15% [3]. It is not unheard of for the plexiform variety to undergo malignant transformation [4].

2. Case Report

A 60 year old lady presented to us at the outpatient department of our institute with a history of difficulty in breathing through her nose and a mass protruding from the right nasal cavity. She gave us a history of the mass being present in her nasal cavity for four years and was slowly growing in size. She first noticed heaviness and difficulty in breathing from the right nasal cavity about four years back and was clinically diagnosed to have a nasal polyp at the

local hospital. Since the mass was growing in size and now was protruding from the nasal cavity, she was referred to medical college hospital for further management.

She gave a history of 3 episodes of epistaxis which were after manipulation. The bleeding was scanty and stopped by itself on application of pressure over the nostril. Patient had mucopurulent nasal discharge which subsided on taking antibiotics. She complained of having generalized weakness and occasional hemicranial headache, which was relieved on taking analgesics. The patient is a known diabetic diagnosed about 12 years back and is on regular medication. She did not give a history of any other abnormal growth or lumps being present over her body or extremities.



Figure 1. Photograph showing mass protruding from the right nasal cavity

On anterior rhinoscopic examination, a pale pinkish polypoidal mass lesion was seen protruding from the right nostril which was soft, non tender, non shrinkable and did not bleed on touch [Figure 1]. A probe could be passed around it for about 1 cm. There was copious amount of mucopurulent discharge in the nasal cavity. The septum was

* Corresponding author:

drbviswanatha@yahoo.co.in (Borlingegowda Viswanatha)

Published online at <http://journal.sapub.org/otolaryn>

Copyright © 2014 Scientific & Academic Publishing. All Rights Reserved

deviated to the left, prominently where the mass was abutting it. Left nasal cavity was normal. There were no palpable neck nodes. On examination her ears and throat were normal.



(a)



(b)

Figure 2. CT scan images showing the extent of the mass in the right nasal cavity

The CT scan showed a mass in the right nasal cavity and extending up to the sphenoid sinus causing pressure symptoms on the septum and pushing it to the left [Figure 2a & 2b]. Heterogeneous contrast enhancement was present. The right maxillary sinus was opaque. It was reported radiographically to be a case of right sinonasal polyposis with features of right maxillary sinusitis.

Accordingly the patient was evaluated for surgical fitness and the mass was excised en masse using endoscopic endonasal approach [Figure 3] under local anesthesia. The mass seemed to arise from the nasal cavity per se and not form the paranasal sinuses. There wasn't much hemorrhage during surgery. The patient tolerated the procedure well and postoperative period was uneventful.

The histopathological examination confirmed the presence of spindle cells with wavy nuclei. Wavy collagen fibres were interspersed with scattered mast cells which confirmed the diagnosis of neurofibroma [Figure 4].



Figure 3. Photograph showing mass excised from the right nasal cavity

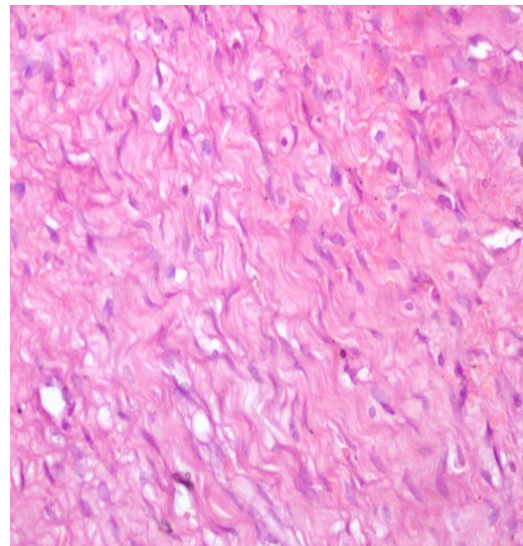


Figure 4. Microphotograph showing spindle cells with wavy nuclei and myxoid stroma

3. Discussion

Neurofibromas are benign peripheral nerve sheath tumors. Nasal cavity involvement is seen in 4% of the cases [1]. On CT scan heterogeneous contrast enhancement is seen in characteristically in neurofibromas [5, 6].

Neurofibromas arise from the non myelinating schwann cells in the peripheral nervous system. They can arise from multiple peripheral nerve sheath tumors which sometimes arise from the head and neck area. Histopathological examination typically shows characteristic curved indented and ovoid schwann cell nuclei with spindle cells and wavy nuclei. Myxoid stroma in wavy fibrils can be appreciated [7, 8]. They are usually associated with neurofibromatosis which are autosomal dominant in nature and have neurofibromas arising from other sites in the body as well. Isolated cases of localised neurofibroma without other associated signs are a rare finding. Specifically, neurofibromas and schwannomas have been reported to

occur in the nasal cavity, ethmoid sinus, maxillary sinus sphenoid sinus and very rarely the frontal sinus [8].

Neurofibromas are more of a histological diagnosis thanks to the characteristic findings like spindle cells with wavy nuclei, wavy collagen fibrils and scattered mast cells. They need to be differentiated from schwannomas and malignant peripheral nerve sheath tumors though. Immunohistochemistry is of use in this regard when neurofibromas show reaction with S-100, NSE and Vimentin [1, 7, 8]. Surgical excision is the mainstay of treatment in symptomatic masses and various methods such as open, endoscopic assisted, endoscopic endonasal approach are employed for the same depending on the site and size of the lesion.

4. Conclusions

A rare case of isolated neurofibroma of the nasal cavity which presented as polypoidal nasal mass was excised en masse using endoscopic methods and was diagnosed histopathologically as neurofibroma of the nasal cavity. The patient was symptom free during 6 months follow-up period. Immunohistochemistry may play an important role in diagnosing nasal neurofibromas. Complete excision of the mass as far as possible provides relief from nasal symptoms. Endoscopic endonasal approach can be used as a treatment modality for such cases.

REFERENCES

- [1] Sudhir B. Sharma L, Hong P, Case report of solitary neurofibroma of the frontal sinus. Case reports in otolaryngology 2012, Article ID 373808, and doi:10.1155/2012/373808.
- [2] Souza L, Oliveiraj, Freitas M, et al. Neurofibromapaciniano: Relato de um casoraro de localizacaoitraoral. Rev Brasil Otorhinolaringol 2003;69:851-54
- [3] Silva V, Agra L, Freitas M, et al. Neurofibroma nasal: Relato de caso e revisao de literature. Rev Brasil Otorhinolaringol 1999;65:172-74.
- [4] Muir D, Neubauer D, Lim IT, Yachnis AT, Wallace MR. Tumorigenic properties of neurofibromin-deficient neurofibroma schwann cells. American Journal of Pathology 2001: 158; 501-513.
- [5] Majoie CB, Hulsmans FJ, Castelijns JA et al. Primary nerve sheath tumors of the trigeminal nerve: clinical and MRI findings. Neuroradiology 1999; 41: 100-108.
- [6] Boedeker CC, Ridder GJ, Kayser G, Schipper J, Maier W. Solitary neurofibroma of the maxillary sinus and pterygopalatine fossa. Otolaryngology and Head and Neck Surgery 2005; 133:458-459.
- [7] Hirao M, Gushiken T, Imokawa H, et al. Solitary neurofibroma of the nasal cavity: resection with endoscopic surgery. J Laryngol Otol 2001; 115: 1012-1014.
- [8] Perzin KH, Panyu H, Wechter S. Nonepithelial tumors of the nasal cavity, paranasal sinuses and nasopharynx- A clinicopathologic study. XII: schwann cell tumors (neurilemoma, neurofibroma, malignant schwannoma). Cancer 1982: 50(10); 2193-2202.