Case Report

Otolaryngology International
Volume 1 Number 2, July - December 2016

Neurofibroma of Nasal Cavity

Angshuman Dutta*, Aditya Bhargava, Deepti Mutreja***, BM Aravind******

Author Affiliation: *Associate Professor, Dept of ENT, Rajiv Gandhi University of Health, Sciences, Jayanagar, Bengaluru.**Professor & Head, Dept of ENT, Command Hospital, Air Force, Bangalore. ***Associate Professor, Dept of Pathology, Command Hospital, Air Force, Bangalore, Resident, Dept of ENT, Command Hospital, Air Force, Bangalore

Reprint Request: Wing Commander (Dr) Angshuman Dutta, Associate Professor, Dept of ENT, Rajiv Gandhi University of Health Sciences, Jayanagar, Bengaluru, Karnataka - 560041.
E-mail: angshumandutta37@gmail.com

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, but isolated neurofibroma occurring in the nasal cavity is an rare entity. Here we report one such case in a 44 yr old man who, presented with a unilateral nasal mass. The mass was histologically confirmed to be a neurofibroma. The clinical presentation, histological features and management of nasal neurofibroma are discussed.

Keywords: Neurofibroma; Schwannoma; Endoscope.

44 yr old male presented with history of mass left nasal cavity for 1 year. On examination there was obliteration of left alar groove. A whitish firm mass filling the nasal cavity was seen which appeared to arise from left lateral nasal wall was sensitive to touch and there was no change in size on vasoconstriction. General examination did not reveal any abnormality.

CECTPNS (Figure 1) showed heterogeneously enhancing mass lesion completely filling the left nasal cavity with dimensions of 14mm*38mm* 59mm. There was opacification of the left maxillary, frontal, ethmoid and sphenoid sinuses. Mass effect was noted in the form of focal indentation and bulge of the nasal septum to the right. No gross bony erosion/sclerosis was seen around the mass. Investigations endoscopic biopsy was suggestive of neurofibroma.

Patient was planned for excision of mass via a lateral rhinotomy incision approach. Flaps were elevated and the left wall of maxilla was exposed. Medial maxillectomy done by taking cuts medial to inferior orbital foramen and along the nasal floor anterior wall of left maxilla was drilled out. Mass was visualised in left maxillary antrum (Figure 2). Mass was carefully dissected from its attachments along the roof and floor of maxillary antrum. Mass was removed in toto after freeing from attachments along ethmoids, sphenoid. Using a 30 degree endoscope through the incision entire cavity was inspected. Small mucoceles were found and drained from posterior ethmoid and sphenoid sinuses. Sphenoidotomy was done and left frontal recess cleared. Left nasal packing was done. Incision was closed in layers. His postoperative recovery was uneventful.

Histopathology showed soft tissue mass 5.5*4.5*1cm (Figure 3) with a smooth and shiny external surface with multiple areas of haemorrhage. Histopathology sections (Figure 4) showed cells are arranged loosely and diffusely composed of spindle shaped cells with wavy comma shaped nuclei scant cytoplasm. The nuclei are seen separated by collagen fibres and myxoid material. No mitotic activity, verocay bodies, palisading of nuclei or hyaline thickening of vessel wall was seen. No atypia/ granuloma seen. Opinion was of Neurofibroma left nasal cavity. On IHC staining S100 was positive.

Patient was kept under regular nasal endoscopic followup for a period of 2 years and no recurrence was noted.

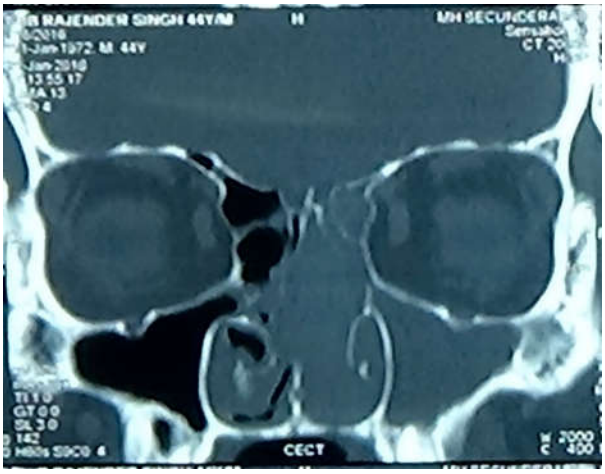

Fig. 1: CT scan PNS showing opacification of left maxillary sinus and nasal cavity

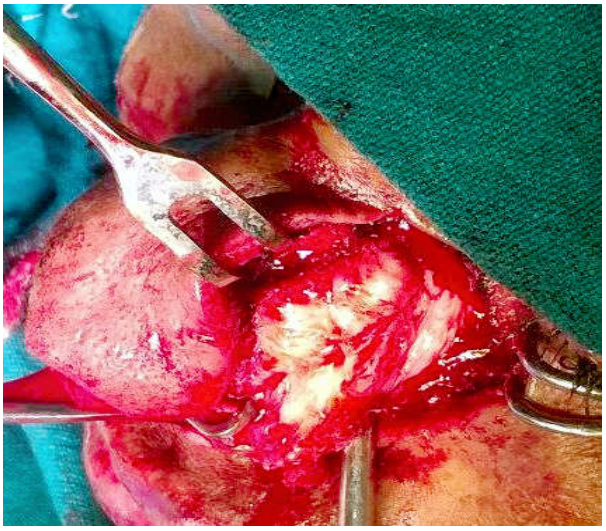

Fig. 2: Exposure of the neurofibroma using the lateral rhinotomy approach

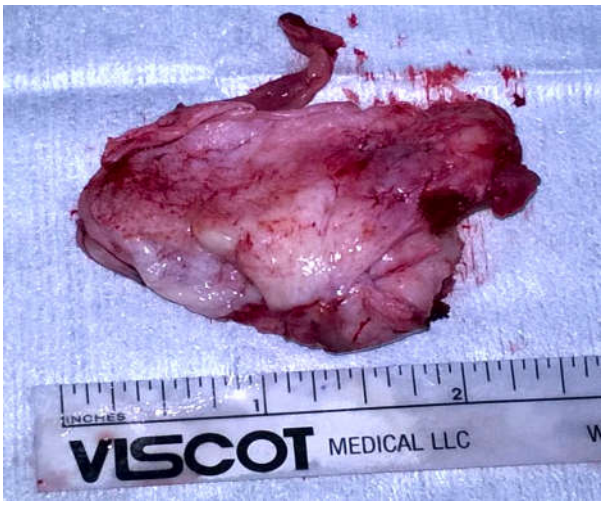

Fig. 3: Photograph of the excised specimen

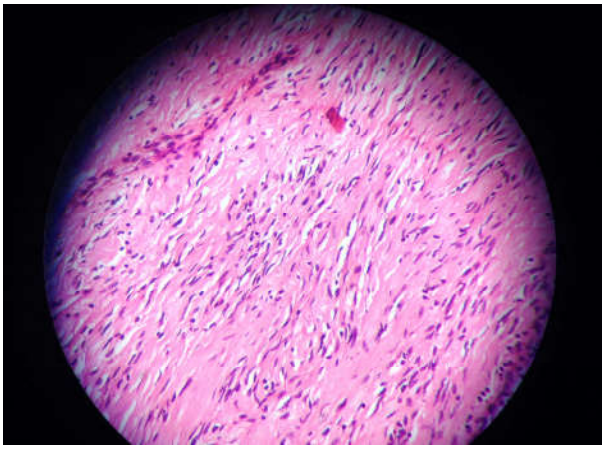

Fig. 4: High power H&E Stain of the neurofibroma showing characteristic spindle cells

Discussion

Neurofibromas are benign peripheral nerve sheath tumors that originate from the nonmyelinating Schwann cells which are derived from neuroectoderm [1]. Neurofibromas can be categorized into dermal and plexiform subtypes. Dermal type is associated with individual peripheral nerves and plexiform types are usually associated with many nerve bundles [1]. The plexiform tumors can rarely undergo malignant transformation [1].

Neurofibromas are usually found in individuals with neurofibromatosis, which is an autosomal dominant condition. There are two types of neurofibromatosis: type 1 (von Recklinghausen disease), which is more common, and type 2, which typically has a more severe course due to central nervous system tumors [2].

Occassionally an isolated neurofibroma can occur without being associated with neurofibromatosis in the gastrointestinal system [3] and very rarely they have been reported to occur in the paranasal sinuses. They have a malignant potential of 2.6% and if associated with von Recklinghausen's disease, the chances for malignant transformation are 3-15%. Neurofibromas commonly occur in third or fourth decades of life.

25% to 45% neurofibromas arise in the head and neck region of which only 4% involve the nasal cavity and paranasal sinuses [4]. Neurofibromas have been reported to occur in the nasal cavity [5], ethmoid sinus, maxillary sinus and sphenoid sinus. In the area of the nose and paranasal sinuses neurofibroma arise from the first and second division of trigeminal nerve and the autonomic plexuses.

Neurofibromas are slow growing benign tumors, however can become very large leading to

compression of local structures, including expansion and erosion of adjacent bone and bone resorption [4]. The clinical features of a neurofibroma depend on the site of the tumor and subsequent involvement of surrounding structures [4]. Imaging for these tumors include CT scanning and magnetic resonance imaging (MRI) to demonstrate the extent and involvement of the tumor. MRI allows better differentiation of the tumor from adjacent soft tissues and better evaluation of any intracranial and intraorbital extension [4] whereas CT scan is useful for bone destruction. The differential diagnosis of neurofibromas include benign tumors of nose like fibroma, papilloma, leiomyoma, and schwannoma [4,5].

The characteristic histologic features of neurofibromas are spindle cells with wavy nuclei, and wavy collagen fibrils. There is a 10% malignant transformation reported in neurofibromas hence it is essential to distinguish it from schwannoma and malignant peripheral nerve sheath tumor [6]. While schwannomas have a capsule; neurofibromas are generally not encapsulated and usually interdigitate with adjacent tissue. In neurofibromas axons are seen to transverse the tumor mass whereas it is not so in schwannomas. Immunohistochemistry is useful in differentiation as neurofibromas show reaction with S-100, NSE Neuron specific enolase and Vimentin [7] but not for desmin or smooth muscle actin which helps in differentiating between neurofibroma and other tumors.

The mainstay of the treatment is complete surgical excision because neurofibromas may infiltrate extensively. The type of approach for removal is dependent on the extent and location of the tumor. Transnasal endoscopic resection can be done if neurofibroma is solitary and located in the nasal cavity and if the origin can be identified. It is important to visualise the origin of the tumor which allows macroscopic complete resection under endoscopy. Open surgical procedures such as lateral rhinotomy, extended ethmoidectomy or partial maxillectomy are required for extensive tumors.

Recurrences are infrequent however rarely tumors with locally invasive tendencies are known to recur after incomplete removal[4]. With a 2-year follow-up, no recurrence was noted for the present case.

Conclusion

A rare case of isolated neurofibroma of the nasal cavity is reported which presented as a unilateral nasal mass and was diagnosed histopathologically as neurofibroma of the nasal cavity. Immunohistochemistry may play an important role in diagnosing nasal neurofibromas.

Conflicts of Interest

The authors have no conflicts of interest to declare

References

1. Muir D., Neubauer D, Lim IT, Yachnis AT, Wallace MR. "Tumorigenic properties of neurofibromin-deficient neurofibroma Schwann cells," American Journal of Pathology. 2001; 158(2):501-513.
2. Goutagny S, Kalamarides M. "Meningiomas and neurofibromatosis," Journal of Neuro-Oncology. 2010; 99(3):341-347.
3. Hindy P, Parvin R, Hanna K, Andrawes S, Gress F, Goodman A. "An isolated neurofibromal polyp of the colon," Case Reports in Gastroenterology. 2012; 6(1):58-62.
4. 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.
5. 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.
6. Yong DK, Chang HB, Jang SS, Kei WS. Transnasal endoscopic excision of an isolated neurofibroma of the nasal septum Rhinology. 1997; 35:89-91.
7. Sharma R, Tyagi I, Banerjee D, Pandey R, "Nasoethmoid schwannoma with intracranial extension. Case report and review of literature," Neurosurgical Review. 1998; 21(1):58–61.