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Case Report

Endoscopic Endonasal Excision of Solitary Neurofibroma Arising from Anterior Nasal Septum: A Case Report and Clinical Review

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Abstract

Background: Neurogenic tumors are rare neoplastic lesions of head and neck region. Benign peripheral nerve sheath tumors arising from nose have been rarely documented in the literature. Only six reports of solitary nasal septal neurofibroma in English literature existed. We present a case of solitary neurofibroma originating from left side of anterior nasal septum in a 40 years old female. Endoscopic endonasal excision was done. At six years follow-up, there was no evidence of recurrence. However rare, solitary neurofibroma must be contemplated in the differentials of unilateral benign tumor involving nasal and paranasal sinuses.

Keywords: Solitary Neurofibroma; Peripheral Nerve Sheath Tumors; Nasal Septum; Endoscopic Excision

Background

Peripheral nerve sheath tumors originate from Schwann and perineural cells [1]. Neurofibroma is a neurogenic tumor that usually presents with Von Recklinghausen's disease. It unlikely presents as a solitary tumor [2,3]. 25-45% of neurofibromas are seen in head and neck region while 4% involves nasal cavity and paranasal sinuses [4]. The treatment of choice for nasal and paranasal sinuses neurofibroma is complete excision [1].

There is limited literature on neurofibroma of the nasal cavity owing to the rarity of this entity. Following six reports by Kim YD., *et al*. [5], Fraczek., *et al*. [6], Kohli PS., *et al*. [7], Kim KS [8], Tall., *et al*. [9], and Katre., *et al*. [10] in English literature, we report a new case having nasal septal as the origin of neurofibroma, exclusively managed by endoscopic endonasal excision. We discuss diagnostic and therapeutic work up of it.

Case Description

A 40 years old female presented with left nasal obstruction for six months, which was gradually progressive, associated with non blood-stained nasal discharge, facial heaviness, mild to moderate intermittent headache and left sided epiphora. On endoscopic examination, there was a nasal mass, with a smooth surface, tender, didn't bleed on touch, impinging on inferior turbinate (Figure 1). All borders except medial were free of any attachment. The computed tomographic scan revealed a heterogeneous soft tissue density filling left nasal cavity extending till posterior choana minimally pushing nasal septum (Figure 2). Endoscopic endonasal excision was done from its anterior nasal septal attachment with a 5 mm margin. The histological examination concluded that the outer surface was irregular and congested. The cut surface was

smooth and grey with focal gelatinous area. Sections showed pseudostratified ciliated columnar epithelium and underlying benign neoplasm comprising of spindle shaped cells in fascicles and whorls. Tumor cells had elongated nuclei with inconspicuous nucleoli (Figure 3). Cells were positive for Vimentin (Figure 4) and S-100, negative for cluster of differentiation (CD)34, CD99 and smooth muscle actin (SMA) suggestive of a benign mesenchymal tumor. At six years follow-up, there was no evidence of recurrence, neither endoscopic nor radiologic.

Figure 1: Zero degree endoscopic examination showing mucosa covered mass in left nasal cavity.

Figure 2: Axial computed tomographic scan showing heterogeneous hypodense mass filling left nasal cavity with posterior extension to choana.

Figure 3: Tumor cells with elongated and inconspicuous nucleoli.

Figure 4: Cells showing positive stain for Vimentin.

Discussion

Peripheral nerve sheath tumors are categorized into neurofibroma, schwannoma, and neurogenic sarcoma. Out of which neurofibroma and schwannoma being benign and neurogenic sarcomas are malignant [1-3]. Follow up of these patients is necessary due to the rare chances of malignant transformation; which is more in neurofibroma as compared to schwannoma. The likelihood increases to 10% when it is associated with neurofibromatosis type 1 (NF 1) [5].

Isolated peripheral nerve-sheath tumors are more common in females in fifth and sixth decades [11]. Neurofibroma of the nasal cavity may be solitary or multiple. If neurofibroma is plexiform,

NF 1 is a common association [12]. In nasal cavity and paranasal sinuses, the tumor arises from trigeminal nerve's first and second division [1]. A combined naso-ethmoid involvement is most common. In order of frequency, it is followed by maxillary sinus, nasal cavity, and sphenoid sinus [1]. In the nasal septum, it is sporadic, and nearly all have been solitary tumors.

Symptoms and signs depend solely on the site of lesion. The usual presentations are nasal obstruction, nasal bleeding, facial pain, swelling, and proptosis. They are primarily centralised in location. The diagnosis is usually made after histopathology. These may be well-circumscribed or intermixed with neighboring connective tissue. Microscopically, they contain spindle cells with fusiform nuclei in connective tissue matrix, which may be myxoid. Immunohistochemically, neurofibroma shows immunoreactivity of S-100 protein, neuron specific enolase and vimentin. It is not reactive for desmin or SMA, which differentiates it from other tumors [5]. Since malignant changes may occur in neurofibroma, the pathologist must differentiate it from neoplasms derived from fibroblasts; fibrous histiocytoma, and other fibro-osseous lesions [13].

Some grow by expansion causing pressure effects while few may show local infiltration. Hence bone erosion with an exception may indicate a non-malignant lesion. High resolution computed tomographic scan can help in delineating the lesion. Magnetic resonance imaging with gadolinium contrast is indicated for exact delineation of tumor and in cases with intraorbital or intracranial extension. Complete surgical resection is the treatment of choice offered for sinonasal neurofibromas. The approach depends on the extent of tumor. Resection should be made after keeping the functional and cosmetic aspects in mind. Neurofibroma is invasive and likely to recur after incomplete removal. Endoscopic endonasal excision is done for solitary and nasal neurofibroma where origin can be identified. However, if it the complete excision is impossible by endoscopic surgery, an open surgical procedure must be undertaken.

An electronic database search on PubMed (MEDLINE), Cochrane Library, Google Scholar, and Embase was made in December 2020 since inception, using terms "nasal septal", "neurofibroma", "solitary". 14 citations were found reporting neurofibroma of the nasal cavity which included twelve case reports [2,5,7-10,14-19] and two case series [6,20] including two and eight patients (Table 1). The site of the lesion were nasal septum in six cases [5-10] and the approach used in all reported cases is mentioned.

S. No.	Year	Authors	Title of the article	Age and gender of patient	Site and Surgical approach
1.	1991	Annino D., *et al.* [2]	A rare cause of nasal obstruction: A solitary NF	69 y/Male	Right posterior nasal cavity/Mid facial devolving with transantral and intranasal approach
2.	1997	Kim YD., *et al.* [5]	Transnasal endoscopic excision of an isolated NF of the nasal septum	58 y/Female	Right septum/Transnasal endoscopic excision
3.	1998	Moreno PM., *et al* [15]	Solitary NF of the inferior nasal turbinate	29 y/Female	Left inferior turbinate/ Endoscopic excision
4.	1999	Chakravarti., *et al.* [16]	Solitary NF causing nasal obstruction	60 y/Male	Not mentioned
5.	2001	Hirao M., *et al.* [14]	Solitary NF of nasal cavity: resection with endoscopic surgery	71 y/Female	Left nasal cavity/Endoscopic excision
6.	2005	Fraczek., *et al.* [6]	Two Cases of nasal NF	68 y/Female 46 y/Female	Right septum/external app Right nasal cavity/Excision with orbital floor resection

7.	2006	Chua N., *et al.* [17]	Solitary nasal NF presenting as compressive optic neuropathy	40 y/Female	Bilateral nasal cavity/Bifrontal craniotomy with transnasal approach
8.	2006	Manganaris., *et al.* [18]	A peripheral nerve sheath tumour as a cause of nasal obstruction	68 y/Female	Right inferior turbinate/ Lateral rhinotomy
9.	2010	Shah S., *et al.* [19]	Neurofibroma of Middle Meatus: A case report and Review of Literature	46 y/Female	Left middle meatus/ Endoscopic sinus surgery
10.	2011	Kohli PS., *et al.* [7]	Endoscopic Excision of Solitary Neurofibroma Arising from Posterior Nasal Septum.	37 y/Female	Left posterior septum/ Endoscopic endonasal excisionsal biopsy
11.	2014	Azani AB., *et al.* [20]	Sinonasal Tract Neurofibroma: A Clinicopathologic Series of 12 Cases with Review of the Literature.	31 y/M 54 y/M 49 y/F 61 y/M 32 y/F 75 y/M 26 y/M 41 y/F	Right nasal cavity/Excision Left nasal cavity/Excision Right nasal cavity/Excision Left nasal cavity/Excision Right nasal cavity/Excision Right nasal cavity/Excision Right nasal cavity/Excision Right nasal cavity/Excision
12.	2015	Kim KS., *et al.* [8]	Solitary Neurofibroma Originating From the Nasal Septum	60 y/Male	Left septum/Excision
13.	2015	Tall A., *et al.* [9]	Solitary Neurofibroma originating from the posterior nasal septum: Transnasal endoscopic resection	48 y/Female	Right posterior septum/ Transnasal endoscopic approach
14.	2017	Katre MI., *et al.* [10]	Neurofibroma of Nasal Cavity and Nasopharynx	17 y/Male	Right posterior septum/ Transnasal endoscopic excision

Table 1: Review of neurofibroma of nasal cavity.

Conclusion

Neurofibroma is a peripheral nerve sheath tumor. Although solitary neurofibroma arising from nasal septum is extremely rare, it should be considered among the differentials of unilateral nasal mass. The tumor is curable by complete excision via an endoscopic endonasal approach, with very little chance of recurrence.

Declarations of Conflicting Interests

The authors declare that there is no conflict of interest.

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