Isolated Plexiform Neurofibroma of the Nasal Tip - A Rare Case Report

Subhadeep Chowdhury[1], Tithi Debnath[2]

[1, 2] Department of Otorhinolaryngology and Head & Neck Surgery, R.G. Kar Medical College, Kolkata, West Bengal, India.

PRESENTATION OF CASE

A 28-year-old female patient presented at ENT OPD with complaints of a slowly growing mass over nasal tip since last 5 years. There were no complaints of pain, nasal obstruction, or epistaxis. There were no skin changes or ulceration over the swelling and sensation was preserved. There was no history of similar kind of swelling in other parts of the body. History of trauma or any form of surgical intervention were excluded. Family history was insignificant. The swelling did not respond to any medications. The patient attended for cosmetic reason solely.

CLINICAL DIAGNOSIS

Clinical examination showed a soft non-tender 4 cm by 4 cm mass over nasal tip and supratip area. Mobility of the mass was restricted and fixed to the alar cartilages. There was no evidence of café au lait spots or any other skin lesions. Anterior rhinoscopy was unremarkable. Diagnostic nasal endoscopy was also performed but no abnormality was detected. CT scan showed non-specific infiltrative subcutaneous lesions. T1W MRI showed an ill-defined hypodense mass over the nasal tip which was abutting the alar cartilages which showed mild enhancement with contrast (figure 1). T2W MRI showed hyperintense and or hypodense central focus (target sign). FNAC was done and report was suggestive of neurogenic tumour, most probably neurofibroma.

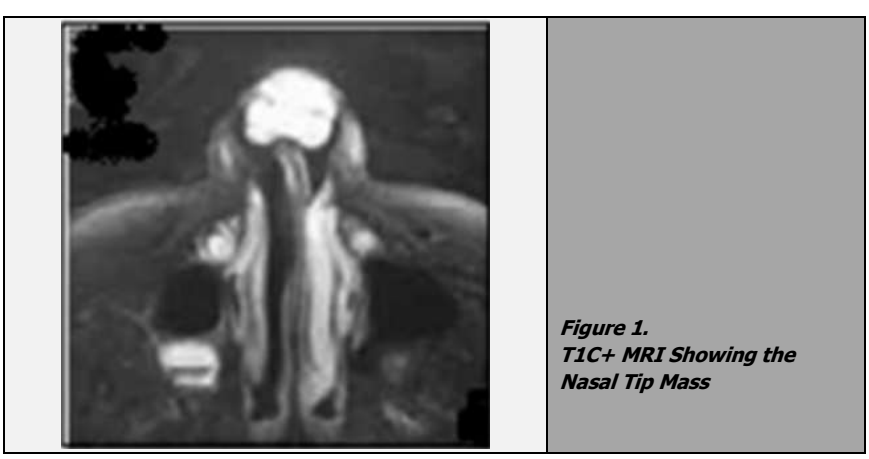

Figure 1.
T1C+ MRI Showing the Nasal Tip Mass

Corresponding Author:
Dr. Tithi Debnath,
Farm Side Road, P.O.
Chinsurah (R.S), Hooghly -712102,
West Bengal. India.
E-mail: tithi_debnath@rediffmail.com

DOI: 10.18410/jebmh/2021/576

How to Cite This Article:
Chowdhury S, Debnath T. IsIsolated plexiform nurofibroma of the nasal tip: a rare case report. J Evid Based Med Healthc 2021;8(33):3169-3171. DOI: 10.18410/jebmh/2021/576

Submission 27-05-2021,
Peer Review 02-06-2021,
Acceptance 28-06-2021,
Published 16-08-2021.

Open rhinoplasty and excision of the mass was done. Intra –operative finding showed a non-capsulated mass which was adherent to the alar cartilages. After meticulous dissection the mass was excised without damaging the underlying alar cartilages. (figure 2 and 3). Immediate post operatively the patient showed improved facial aesthetics. (figure 4). Post- operative specimen shown in figure 5.

Figure 2. Showing Dissection of the Mass From Underlying ALAR Cartilage

Figure 3. Mass after Dissection

Figure 4. Showing Post-Operative Status

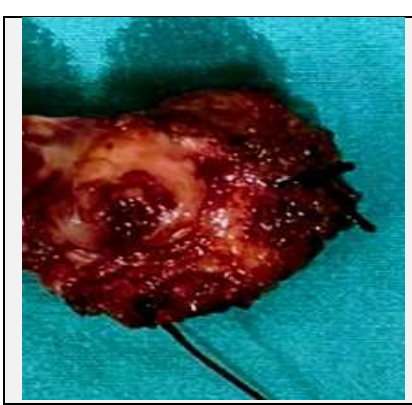

Figure 5. Excised Specimen

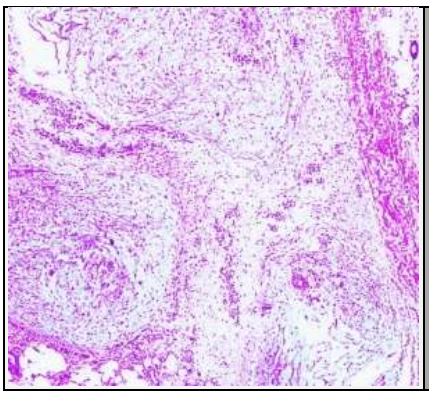

Figure 6. Histopathology Picture

DIFFERENTIAL DIAGNOSIS

- Nasal tip Dermoid
- Lipoma
- Leiomyoma
- Fibrous papule
- Rhinophyma
- Haemangioma
- Facial eosinophilic granuloma
- Neurofibroma
- Neurilemmoma
- Melanoma
- Basal cell carcinoma
- Squamous call carcinoma
- Kaposi's sarcoma

PATHOLOGICAL DISCUSSION

Neurofibromatosis is a clinical entity which is associated with development of neoplasm from the brain, spinal cord or other peripheral nerves.[1] It is of three types mainly of which type 1 also known as Von Recklinghausen's disease which is associated with skin lesions and growth of tumours along nerves in skin, brain and also other parts of the body.[2] Neurofibromas are localized mostly benign peripheral nerve sheath tumours arising from proliferation of axons, Schwann cells, perineural cells and endoneurium. They can occur either sporadic or as a part of autosomal dominant Neurofibromatosis type 1.[3] Isolated neurofibroma of the nasal tip are rare. Also their treatment are challenging in view of their position and high recurrence rate. The first reported cases of external nasal neurogenic tumours were reported by New and Devine in 1947[4] and Das Gupta et al. in 1969.[5] In 2007 Rameh et al. published an article claiming to report the first case of solitary plexiform neurofibroma of the nasal tip.[6]

Nose being at the center of the face plays a significant role in facial beauty.[7] Any lesion involving nose is presented early by the patients. However, our patient presented late as she was trying different medications so that the swelling dissolved without surgery instead it gradually increased in size which prompted her to visit ENT OPD presently. The lesion came out to be neurofibroma which was unthought of as an isolated neurofibroma without other associated

symptoms of Neurofibromatosis is extremely rare that to over the nasal tip.[6] Though by imaging and FNAC, it was assumed to be a neurogenic tumour but only radiology is not enough to prove it[8] and hence definitive diagnosis was only obtained by histopathology and immunohistochemistry. A high local recurrence rate is very common in these cases as these are aesthetically demanding and hence to maintain alar cartilage integrity often these lesions are incompletely excised.[9] This has to be counseled to the patient beforehand.

DISCUSSION OF MANAGEMENT

Treatment of the nasal lesions are challenging because of their position, high recurrence and more importantly cosmesis.[10] Nasal tip lesions are more difficult because skin here is more thick, more sebaceous and more adherent than the nasal dorsum and hence nose can be divided into an anatomical and an aesthetic subunit and nasal tip falls under the latter.[11,12] Treatment varies upon various factors like site and size of the lesion, mobility, benign or malignant, recurrence rate, expertise and patients expectations.[13,14] In our case the lesion was big and thick so leaving an excess skin after excision.

Rameh et al reported a case of solitary plexiform neurofibroma which was treated by open rhinoplasty approach.[6] A recently published study by Rao et al reported another such case which was treated by open rhinoplasty and alar cartilages were preserved but the patient was lost to follow up.8 In the literature there have been six documented cases of solitary nasal tip neurogenic tumors of which three were Neurilemmoma, two solitary plexiform neurofibroma and another one was neurogenic tumour not otherwise specified[(NOS).[6] In all cases the surgical excision was the primary modality of treatment and the surgery performed was open rhinoplasty. Though other surgical modalities like Mohs micrographic surgery is an useful technique for difficult to treat nasal parts which pose an aesthetic and or functional challenge.[9] In our case too we went by following the literature and excised the mass by open rhinoplasty approach.

FINAL DIAGNOSIS

Plexiform neurofibroma of the nasal tip

CONCLUSIONS

Sinonasal neuroendocrine tumours are rare and as Plexiform neurofibroma is non- encapsulated and diffuse and also being in a cosmetically sensitive area hence surgically challenging so there is increased chances of local recurrence. As rate of malignant transformation is low in these cases aggressive disfiguring incisions are not required.

Financial or other competing interests: None.
Disclosure forms provided by the authors are available with the full text of this article at jebmh.com.

REFERENCES

[1] Kim KS. Complete removal of external nasal neurofibroma by endonasal approach. The Journal of Craniofacial Surgery 2015;26(7):e562-564.
[2] Ferner RE, O'Doherty MJ. Neurofibroma and schwannoma. Current Opinion in Neurology 2000;15(6):679-684.
[3] Evans HL. Sporadic superficial diffuse neurofibromas with repeated local recurrence over many years and a tendency toward malignant change: a report of 3 cases. Am J Surg Pathol 2013;37(7):987-994.
[4] New GB, Devine KD. Neurogenic tumors of nose and throat. Arch of Otolaryngol 1947;46(2):163-179.
[5] DasGupta TK, Brasfield RD, Strong EW, et al. Benign solitary schwannoma (Neurilemomas). Cancer 1969;24(2):355-366.
[6] Rameh C, Husseini S, Tawil A, et al. Solitary plexiform neurofibroma of the nasal tip: case report and review of literature. International Journal of Pediatric Otorhinolaryngology Extra 2007;2(2):116-119.
[7] Amodeo CA. The central role of the nose in the face and the psyche: review of the nose and the psyche. Aaesthet Plast Surg 2007;31(4):406-410.
[8] Rao AG, Lakhsmi TS, Haritha S. Diffuse Neurofibroma. Indian J Dermatol Venereol Leprol 2003;69:17-18.
[9] Trieu D, Drosou A, Schwartz MR, et al. Myxoid neurofibroma treated with mohs micrographic surgery letters and communications. American Society for Dermatologic Surgery 2015;41(1):166-168.
[10] Hirao M, Gushiken T, Imokawa H, et al. Solitary neurofibroma of the nasal cavity: resection with endoscopic surgery. J Laryngol Otol 2001;115(12):1012-1014.
[11] Venkata N, Mohan J, Kartik P, et al. Plexiform neurofibroma of nasal tip. International Journal of Otolaryngology and Head & Neck Surgery 2015;4(6):423-428.
[12] Cook JL. Nasal reconstruction in flaps and grafts in dermatologic surgery. New York, NY: Elsevier 2008: p. 191-215.
[13] Vimal C, Rao RS, Sekhar G, et al. Recurrent diffuse neurofibroma of nose associated with neurofibromatosis type 1: a rare case report with review of literature. Indian Journal of Dermatology 2015;60(6):573-577.
[14] Fattahi TT. An overview of facial aesthetic units. J Oral Maxillofac Surg 2003;61(10):1207-1211.