

## A RARE CASE OF SOLITARY NEUROFIBROMA OF NASAL TIP AND COLUMELLA OF NOSE

**Authors:** Mohit Lavania (1), Gunjan Rawa\* (2).

**Authors Affiliations:** (1) Head Of Department of Otolaryngology Head and Neck Surgery – Chandramohan Singh Negi Base Government Combined Hospital, Kotdwara, Distt- Pauri Garhwal, Uttarakhand. (2) Dr. Gunjan Rawat(\*) - Oral and Maxillofacial surgeon- Kotdwara, Distt-Pauri Garhwal, Uttarakhand.

### ABSTRACT

#### INTRODUCTION

Neurofibroma is a most common benign nerve sheath tumor arising in peripheral nervous system. Neurofibroma are lesions arising from proliferation of axons, schwann cells, fibroblasts, perineural cells and endoneurium. Localized neurofibroma occur as solitary, superficial cutaneous tumour in individuals who do not have Neurofibromatosis 1 (NF1). Localised Neurofibroma of external nose is rare lesion. Treatment of which is challenging in view of their position, cosmesis, reoccurrence rate. We report a case of neurofibroma involving unusual site of face, which are subcutaneous tissue of apex and columella of nose, and present a brief review of literature.

#### CONCLUSION

Neurofibroma is a peripheral nerve-sheath tumour. Neurofibroma of the nose is extremely rare, and curable by complete local excision.

**KEYWORDS:** Neurofibroma, Benign, Solitary, Peripheral Nerve Tumors, Nose.

#### INTRODUCTION

The clinical appearance of the peripheral nerve sheath tumors is of soft tissue mass which include involvement of neural structures. The benign nerve sheath tumors are more common than malignant tumors. Benign peripheral nerve sheath tumors are divided into entities namely: localized neurofibroma and schwannoma.<sup>1</sup> These

lesions are believed to arise from schwann cells. localized neurofibroma is slow growing, well circumscribed, but not encapsulated lesion like schwannoma. Localized neurofibroma present as solitary tumour and are curable by adequate primary excision. Most commonly they are found in the head, neck and flexoral surfaces of the upper and lower extremities (stout, 1935).<sup>2</sup> Neurofibroma are less common than schwannoma (neurilemoma, neurinoma). Both tumours are encountered as isolated lesions in the general population, but multiple neurofibromas are characteristic of neurofibromatosis type 1 (NF1) whereas bilateral vestibular schwannomas are indicative of neurofibromatosis type 2 (NF2).<sup>3</sup>

With a frequency of ~ 5% of soft tissue tumors, localized neurofibroma is usually observed in younger individuals, particularly between 20 and 30 years of age. Most lesions originate from small nerves and therefore occur in superficial locations. Deep-seated tumors are seen with neurofibromas originating from major nerves.<sup>1</sup> Vestibular nerve is the most common origin of these tumors in the head and neck region.<sup>4</sup> In this article we report a case of solitary neurofibroma of nose which was successfully excised surgically.

#### CASE REPORT

A female patient aged 30 years presented to the Department of Otorhinolaryngology, with a chief

complaint of a slow and progressively growing soft tissue mass involving the tip and columella of the nose of several years duration. Patient did not complain of facial pain, headache, epistaxis, nasal discharge, or adenoid. There was no history of trauma or previous surgical manipulation in this area. Family history was negative for any similar conditions. Patient had no history of systemic diseases in the past. Neurologic examination of the patient was normal. There was no lymphadenopathy in the neck. There was no pigmentation and mass on the skin of other parts of the body.

On physical examination, there was approximate a 2.5 cm × 2cm soft tissue mass involving the tip and columella region of the nose(fig.1). The mass was non-tender, with no overlying skin changes. The osteocartilaginous framework of nose appeared normal with partial obliteration of the anterior nares.



**Fig1.** Soft tissue mass involving tip of nose.



**Fig2.** Partial obliteration of anterior nares.



**Fig3.** Excision of mass



**Fig4.** Suture placement

The mass was biopsied. Pathology was consistent with a benign neurogenic lesion, suspicious of either a neuroma or a neurofibroma. Complete excision of the mass was then performed via an open rhinoplasty approach(fig.3,4). Final Histopathology revealed a skin covered tissue piece with unremarkable epidermis. Underlying dermis shows a tumor composed of proliferation of spindle shaped to wavy nuclei with interspersed collagen bundles. Interspersed nerve bundle are appreciated. No verrocay bodies, no nuclear palisading appreciated. No areas of Necrosis/ brisk mitosis or atypia seen, suggestive of neurofibroma of nose.

#### **DISCUSSION**

Neurofibromas are benign, heterogeneous peripheral nerve sheath tumours arising from the connective tissue of peripheral nerve sheaths,

---

---

especially the endoneurium. Schwannomas are benign encapsulated tumours originating from the Schwann cells of the peripheral nervous system. Both tumours are encountered as isolated lesions in the general population, but multiple neurofibromas are characteristic of neurofibromatosis type 1 (NF1) whereas bilateral vestibular Schwannomas are indicative of neurofibromatosis type 2 (NF2).<sup>3,6</sup>

Neurofibromatosis (NF), a disease described in 1882 by Friedrich Daniel Von Recklinghausen, is a neuro ectodermal abnormality constituted by a set of clinical symptoms that compromise the skin, nervous system, bones, eyes and other sites. Symptoms manifest differently in each patient, even those within the same family, with a highly variable expression.<sup>5</sup>

Neurofibromas are benign peripheral nerve sheath tumors composed of a mixed population of Schwann cells, perineurial-like cells, and fibroblasts present with interspersed nonneoplastic nerve fibers, collagen fibers, and myxoid matrix.<sup>6,7</sup> These lesions appear as soft, skin-colored papules or small subcutaneous nodules.

Neurofibromas has subtyped based on their anatomic localization and gross appearance as cutaneous neurofibroma (localized or diffuse), intraneural neurofibroma (localized), plexiform neurofibroma, or massive soft tissue/visceral neurofibroma.<sup>7</sup> The majority of neurofibromas are sporadically-occurring and localized and have an extremely low risk of malignant transformation. The plexiform type is pathognomonic for neurofibromatosis type 1 (NF 1). It carries an increased risk of malignant transformation.<sup>6</sup> Other classification system assigns 1 of 5 subcategories to describe the clinical appearance of cutaneous neurofibroma nascent, flat, sessile, globular, or pedunculated.<sup>8</sup>

Neurofibromas are typically grayish tan in color and range in consistency from gelatinous to firm. Cutaneous neurofibromas can be nodular/globular and well circumscribed (localized) or can diffusely involve subcutaneous tissue and skin (diffuse cutaneous neurofibroma). Massive soft tissue neurofibromas are large diffuse tumors that often cause localized enlargement of a single limb or regional soft tissue (formerly termed elephantiasis neuromatosa)<sup>7</sup>

In the head-and-neck region, Peripheral nerve sheath tumors are believed to arise from schwann cells of sensory nerve fibres. They often originates from the vestibular nerve. However, neurofibroma arising from the nose and paranasal sinuses are rare<sup>10,11</sup>. When they are present in this area, a combined nasal-ethmoid involvement is the most common, followed by maxillary sinus, intranasal, and sphenoid sinus.<sup>11</sup> Neurofibroma those from the nasal septum are also extremely rare<sup>11,12</sup>

The great majority of the peripheral nerves sheath tumours reported were schwannomas in nose and paranasal sinuses then neurofibroma. Neurofibroma can rarely occur as a solitary entity, mostly multiple in association with von Recklinghausen's disease (NF1). Isolated neurofibroma in head and neck region are more common in females, with fifth and sixth decades.<sup>10,11</sup> In the nose and paranasal sinuses, the tumour arises from the first and second division of the trigeminal nerve and from autonomic plexuses, but cannot arise from the olfactory nerve, which has no Schwann cells. if neurofibroma is present as a small solitary tumour, it is curable by adequate primary excision.<sup>11,12</sup>

Approximately 90% of cases occur sporadically, while the remaining cases are associated with NF1 or NF2.<sup>6, 13</sup> The prevalence of neurofibromatosis is 1:2,500- 1:3,000 live births.

---

---

According to National Institute of Health (NIH), 1988, Clinical Diagnostic criteria for NF1 is

1. Six or more cafe-au-lait spots greater than 5 mm prepubertal and greater than 15 mm post-pubertal.
2. Presence of axillary and/or inguinal ephelides.
3. Two or more cutaneous neurofibromas or one plexiform neurofibroma.
4. Optic pathway glioma.
5. Two or more Lisch nodules.
6. Bone lesion suggestive of NF1: sphenoid dysplasia, dysplasia or thinning of the cortex of long bones with or without pseudo - arthrosis.
7. A first-degree relative with definite NF1.

Atleast Cafe-au-lait spots (MCCL), Ephelid (Crowe's sign), and Cutaneous neurofibromas (Button holing sign) are of special relevance, they constitute 3 of the 7 NIH clinical diagnostic criteria.<sup>13</sup>

The NF1 gene (tumor suppressor gene) which codes for protein neurofibromin is located in pericentromeric region of chromosome 17. Neurofibromin is expressed in Schwann cells, leukocytes, melanocytes, adrenal gland and other tissue of central nervous system. Neurofibromin is a tumor suppressor that functions in the RAS/MAPK and mTOR pathways. Mosaicism can occur, resulting in the segmental, generalized, or gonadal NF1 gene. Spontaneous mutation of NF1 gene that controls production of non functional neurofibromin, that cannot regulate cell growth and division. This leads to formation of neurofibroma along nerve fibres. In about 50% of cases altered gene is inherited, while others result from new mutations.<sup>13,14</sup>

NF2 is a dominantly inherited tumour prone disorder characterised by the development of multiple Schwannomas and meningiomas. NF2 shows loss of constitutional heterozygosity of chromosome 22, with DNA markers lost in

tumours. NF2 gene codes for merlin which is a cell membrane protein that is also a tumor suppressor that functions in the PI3kinase/Akt, Raf/MEK/ERK, and mTOR pathways.<sup>15</sup>

The Diagnostic criteria according to National Institute of Health (NIH) for NF2 :Bilateral vestibular schwannomas (VS) or family history of NF2 plus Unilateral, any two of: meningioma, glioma, neurofibroma, schwannoma, and posterior subcapsular opacities. With additional criteria Unilateral VS or Any two entity: meningioma, glioma, neurofibroma, schwannoma.<sup>14,15</sup>

Histologically, Neurofibroma shows relatively hypocellular proliferation of bland, pale eosinophilic spindle cells with rather wavy, S-shaped or buckled nuclei set in a copious fibrillary or rather myxoid background. Usually, mitoses are not seen. Small nerve fibers are usually readily identified within the tumour. The stroma of these lesions occasionally undergoes marked myxoid or hyaline change. Ultrastructurally, this is composed of an admixture of Schwann cells and perineural fibroblasts, and these cells are S-100 positive. In contrast, a schwannoma is a truly encapsulated tumour and shows two distinct patterns, Antoni-A and Antoni-B. The nuclei are arranged in palisades with space between the rows, forming the so-called Verocay bodies.<sup>11</sup>

Usually common symptoms and signs are nasal obstruction, epistaxis, facial pain, swelling, and proptosis. neurofibroma are asymptomatic and primarily centripetally located. The diagnosis of neurofibroma is usually made only when the lesion is biopsied, because of the non-specific nature of these symptoms and signs.

The treatment of choice for benign nerve sheath tumor is complete surgical excision. Neurofibroma are invasive tumors than



---

schwannoma and more likely to reoccur after removal. Extensive surgical resection is usually recommended which has shown good prognosis. Malignant transformation is rare, unless patient is having neurofibromatosis.<sup>2,10,11</sup>

Plexiform neurofibromas have malignant potential. There 13% risk for plexiform neurofibromas to develop into malignant peripheral nerve sheath tumors. Imatinib has been shown to decrease plexiform neurofibroma size.<sup>6</sup>

Ketotifen target mast cells H1 histamine receptor and decrease symptoms of pain, itch. Other drugs like Everolimus and bevacizumab target, mTor, VEGF showed Minimal changes in cNF. A Topical application of 5% imiquimod, which is an immune-response modifier that acts as a toll-like receptor 7 (tlr-7) after 4 months, cNF showed a 15% reduction in tumor volume, imiquimod showed minimal changes for cutaneous neurofibroma. Co2 laser is effective, but has high risk of scarring hypopigmentation or hyperpigmentation that alter cosmetic outcomes. Surgical removal has been the mainstay of therapy.<sup>16,17</sup>

## CONCLUSION

Neurofibroma is a peripheral nerve-sheath tumour. Solitary Cutaneous neurofibroma of the nose is extremely rare, and curable by complete local excision via a open rhinoplasty approach.

## DECLARATION

Ethics approval and consent to participate: The study was approved by Institutional Ethics committee.

**Conflict of Interests-** The authors declares that there are no conflicts of interest.

**Funding:** The study was self funded

**Acknowledgement:** None

## REFERENCES

1. Woertler K. Tumors and tumor-like lesions of peripheral nerves. *Semin Musculoskelet Radiol.* 2010 Nov;<sup>14</sup>(5):547-58. doi: 10.1055/s-0030-1268073. Epub 2010 Nov 11. PMID: 21072731.
2. Stevens DJ, Kirkham N. Neurofibromas of the paranasal sinuses. *J Laryngol Otol.* 1988 Mar;<sup>102</sup>(3):256-9. doi: 10.1017/s0022215100104669. PMID: 3356936.
3. Ferner RE, O'Doherty MJ. Neurofibroma and schwannoma. *Curr Opin Neurol.* 2002 Dec;<sup>15</sup>(6):679-84. doi: 10.1097/01.wco.0000044763.39452.aa. PMID:12447105.
4. Sakaguchi Y, Ikeda O, Ohgaki K, Oki E, Chinen Y, Sakamoto Y, Minami K, Toh Y, Okamura T. Totally Laparoscopic Gastrectomy for Gastric Cancer Associated with Recklinghausen's Disease. *Diagn Ther Endosc.* 2010;2010:682401. doi: 10.1155/2010/682401. Epub 2010 Jun 23. PMID: 20672006; PMCID: PMC2905906.
5. Antônio JR, Goloni-Bertollo EM, Trídico LA. Neurofibromatosis: chronological history and current issues. *An Bras Dermatol.* 2013 May-Jun;<sup>88</sup>(3):329-43. doi: 10.1590/abd1806-4841.20132125. PMID: 23793209; PMCID: PMC3754363.
6. Messersmith L, Krauland K. Neurofibroma. In: *StatPearls. Treasure Island (FL): StatPearls Publishing; 2023 Jan.*
7. Skovronsky DM, Oberholtzer JC. Pathologic classification of peripheral nerve tumors. *Neurosurg Clin N Am.* 2004 Apr;<sup>15</sup>(2):157-66. doi: 10.1016/j.nec.2004.02.005. PMID: 15177315.
8. Ortonne N, Wolkenstein P, Blakeley JO, Korf B, Plotkin SR, Riccardi VM, Miller DC, Huson S, Peltonen J, Rosenberg A, Carroll SL, Verma SK, Mautner V, Upadhyaya M, Stemmer-

- Rachamimov A. Cutaneous neurofibromas: Current clinical and pathologic issues. *Neurology*. 2018 Jul 10;91(2 Suppl 1):S5-S13. doi: 10.1212/WNL.00000000000005792. PMID: 29987130.
9. Kleihues P, Louis DN, Scheithauer BW, Rorke LB, Reifenberger G, Burger PC, Cavenee WK. The WHO classification of tumors of the nervous system. *J Neuropathol Exp Neurol*. 2002 Mar;61(3):215-25; discussion 226-9. doi: 10.1093/jnen/61.3.215. PMID: 11895036.
  10. Kim YD, Bai CH, Suh JS, Song KW. Transnasal endoscopic excision of an isolated neurofibroma of the nasal septum. *Rhinology*. 1997 Jun;35(2):89-91. PMID: 9299658.
  11. Annino DJ Jr, Domanowski GF, Vaughan CW. A rare cause of nasal obstruction: a solitary neurofibroma. *Otolaryngol Head Neck Surg*. 1991 Apr;104(4):484-8. doi: 10.1177/019459989110400410. PMID: 1903860.
  12. Hillstrom RP, Zarbo RJ, Jacobs JR. Nerve sheath tumors of the paranasal sinuses: Electron microscopy and histopathologic diagnosis. *Otolaryngology–Head and Neck Surgery*. 1990;102(3):257-263. doi:[10.1177/019459989010200309](https://doi.org/10.1177/019459989010200309)
  13. Hernández-Martín A, Duat-Rodríguez A. An Update on Neurofibromatosis Type 1: Not Just Café-au-Lait Spots, Freckling, and Neurofibromas. An Update. Part I. Dermatological Clinical Criteria Diagnostic of the Disease. *Actas Dermosifiliogr*. 2016 Jul-Aug;107(6):454-64. English, Spanish. doi: 10.1016/j.ad.2016.01.004. Epub 2016 Mar 12. PMID: 26979265.
  14. Barker D, Wright E, Nguyen K, Cannon L, Fain P, Goldgar D, Bishop DT, Carey J, Baty B, Kivlin J, et al. Gene for von Recklinghausen neurofibromatosis is in the pericentromeric region of chromosome 17. *Science*. 1987 May 29;236(4805):1100-2. doi: 10.1126/science.3107130. PMID: 3107130.
  15. Evans DG. Neurofibromatosis type 2 (NF2): a clinical and molecular review. *Orphanet J Rare Dis*. 2009 Jun 19;4:16. doi: 10.1186/1750-1172-4-16. PMID: 19545378; PMCID: PMC2708144.
  16. Chamseddin BH, Le LQ. Management of cutaneous neurofibroma: current therapy and future directions. *Neurooncol Adv*. 2019 Dec 16;2(Suppl 1):i107-i116. doi: 10.1093/noajnl/vdz034. PMID: 32642736; PMCID: PMC7317049.
  17. Riccardi VM. Mast-cell stabilization to decrease neurofibroma growth. Preliminary experience with ketotifen. *Arch Dermatol*. 1987;123(8):1011–1016.

Copyright: © 2023 Lavania M, et al. This is an open-access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium by the author.

**\*Corresponding author:**

Dr. Gunjan Rawat  
 Oral and Maxillofacial Surgeon,  
 Kotdwara, Distt-Pauri Garhwal, Uttarakhand.  
 E-mail: [rawatgunjan23@gmail.com](mailto:rawatgunjan23@gmail.com) / [mohit.lavania@gmail.com](mailto:mohit.lavania@gmail.com)  
 ORCID ID number: 0000-0003-0068-7263  
 Phone Number: 7078945401, 8958945403

**How to cite this article**

Lavania M, et al - A Rare Case Of Solitary Neurofibroma Of Nasal Tip Andcolumella Of Nose -UPJOHNS; Dec. 23; 11(2); page: 54-59  
 DOI: <http://doi.org/10.36611/upjohns/volume11/Issue2/9>  
 Orchid Id: <https://orcid.org/0000-0001-9316-8678>



This work is licensed under a Creative Commons Attribution 4.0 International License  
 Copyright © 2020 –UPJOHNS