

## **Trouble at the tip: A case report on Neurofibroma of the nasal tip and review of the literature**

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## Abstract

Neurofibroma are localised, benign peripheral nerve sheath tumours arising from proliferation of axons, Schwann cells, fibroblasts, perineural cells and endoneurium. Isolated neurofibroma of the nasal tip are rare and their treatment are challenging in view of their position, high recurrence rate and cosmesis. We report a case of a diffuse neurofibroma of the nasal tip in a 39-year-old female and discuss challenges encountered in the management of such cases and issue recommendations in treatment planning.

## Introduction

Neurofibroma are localised, benign peripheral nerve sheath tumours arising from proliferation of axons, Schwann cells, fibroblasts, perineural cells and endoneurium.<sup>1,2,3</sup> They can be sporadic or be part of the autosomal dominant disorder Neurofibromatosis Type 1, also known as Von Recklinghausen's disease.<sup>4</sup> They can be typical or diffuse, the latter being an unusual variant with the skin and subcutaneous tissue diffusely infiltrated.

Isolated neurofibroma of the nasal tip are rare and their treatment is challenging in view of their position, high recurrence rate and cosmesis.

We report a case of a diffuse neurofibroma of the nasal tip in a 39-year-old female.

A 39-year-old lady was referred to the ENT clinic with a lesion on the tip of her nose, clinically thought to be a lipoma, which had been slowly increasing in size since childhood. She had no other nasal symptoms and no previous history of trauma or surgery to the nose. She was a known case of Neurofibromatosis Type 1 albeit with minimal clinical manifestations. On examination, there was a soft mass at the nasal tip situated mostly over the right lower lateral cartilage and over the right nasolabial fold with a diameter of about 2.5 cm (Figure 1). Endoscopic nasal examination was normal.

The patient underwent excision of the nasal mass via an open rhinoplasty approach. The mass was dissected from overlying skin and underlying cartilage and sent for histology. The alar cartilages were found to be very soft, with the left cartilage being smaller than the right.

## Case Report

Considerable excess skin remained (Figure 2).

The lesion was confirmed to be a diffuse neurofibroma (incompletely excised), with immunohistochemistry showing widespread expression of S100 protein and CD34 with no lesional expression of EMA or neurofilament protein.

At 9 months post-op, the nasal tip remains rather bulbous due to the excess skin with poor tip support and definition (Figure 3). The patient has thus been listed for revision surgery to reduce the bulbous tip and create a dorsum and nasal columella, and reconstruct the lateral alar cartilages using a conchal cartilage graft.

### Discussion

Patients with nasal lesions tend to present early and are commonly self-referred, due to the exposed and highly visible location of the nose<sup>5</sup>. Our patient presented quite late and this resulted in gradual loss of the cartilaginous support of the tip.

Treatment of nasal tip lesions is challenging in view of their position, high recurrence rate and cosmesis. The nose can be divided into anatomical and anaesthetic subunits.<sup>5</sup> The nasal tip, which is part of the aesthetic tip subunit poses an even bigger challenge as the skin here is thicker, more sebaceous,

more adherent and less flexible when compared to the anatomical dorsum subunit.<sup>5-10</sup> Treatment is varied and depends upon multiple factors namely, site and size of the lesion, whether the lesion is benign or malignant, the recurrence rate, local expertise and patients' preference. In our case, the lesion was large, leaving considerable excess skin, which was rather thick. The patient was not specifically consented for skin excision, so we could not proceed with trimming excess skin. In fact, post-operatively, the nasal tip still looked rather bulbous. On the other hand, delaying skin resection may in turn result in natural contraction, possibly obviating the need for a second stage procedure. In this particular case however, the tip lacked cartilaginous support, thus further reconstruction was still deemed necessary.

The lesion turned out to be a neurofibroma, which was not expected. Preoperative imaging might have pre-empted this as radiologically neurofibromas and schwannomas have a high signal on T2-weighted sequences, however a diagnosis cannot be reached on imaging alone.<sup>11</sup>

A high local recurrence rate is typical with incomplete surgical excision.<sup>4,12</sup> The patient needs to be made aware of this as it might affect patient choice as regarding

the extent of reconstruction he/she are willing to invest in. Also, rarely, neurofibromas can undergo malignant transformation.<sup>4, 13</sup>

Rameh et al. in 2007 reported a case with a solitary plexiform neurofibroma of the nasal tip, which was treated by an open rhinoplasty approach. The patient in question was a teenager and thus maximal attention was exerted to preserve the cartilaginous structure so as not to interfere with the patient's normal growth and development. The open rhinoplasty approach allowed the author to successfully excise the lesion whilst preserving the upper and lower lateral alar cartilages.<sup>3</sup>

A recently published case report by Nataraj et al. reported another nasal tip neurofibroma managed with open rhinoplasty approach with preservation of alar cartilages. The patient was unfortunately lost to follow-up so long-term outcomes are not known.<sup>11</sup>

In the literature there have been six reported cases of solitary nasal tip neurogenic tumours, of which three were schwannomas, two were solitary plexiform neurofibroma, and another one was neurogenic tumour not otherwise specified (NOS).<sup>3</sup>

In all documented case reports, surgical excision was the primary modality of treatment, including midline dorsal

incisions, inter-cartilagenous and butterfly incisions.<sup>3</sup>

Other treatment modalities for management of neurofibromata in other anatomical subunits include Mohs micrographic surgery, which was used for a patient with a myxoid neurofibroma who had had two recurrences after two surgical excisions for a neurofibroma on the nasal dorsum, proximal to the tip.<sup>14</sup> Mohs micrographic surgery is a useful technique for difficult-to-treat nasal subunits which pose a cosmetic and/or functional challenge.<sup>14</sup>

### **Conclusion**

An open rhinoplasty approach provides good access for excision of nasal tip lesion and subsequent reconstruction. In long-standing lesion large lesions, consider the possibility of loss of support due to softening of cartilage and make provisions to reconstruct and provide additional tip support, possibly by the use of grafts.

Excess skin can be an issue when large lesions are removed – this should be taken in to account when planning surgery and consenting.

Pre-operative radiography may aid diagnosis.

Neurofibroma of the nasal tip are likely to be incompletely excised due to this being a cosmetically-sensitive area, hence they

can have a high recurrence rate. The latter might affect the decision as regards the extent of reconstruction preferred by both surgeon and patient.

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**Figure Legends**

Figure 1. Pre-operative photos.



Figure 3. Appearance five months post-op.

Figure 2. Intra-operative photo following removal of the nasal tip neurofibroma, external rhinoplasty approach and excess skin shown.



