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(CASE REPORT)

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Neurofibroma unusual site, the nasal tip: Case report

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Abstract

Neurofibromas are considered as benign tumors of the peripheral nerve sheath which originates from the proliferation of elements of the peripheral nerve including; Schwann cells, axons, fibroblasts, perineurial and endoneurial cells. Neurofibroma can arise in any anatomic location, however having a neurofibroma originating from the external nose more specifically the nasal tip is extremely rare. In this article we are presenting a rare case of nasal tip neurofibroma in 11 years old female child.

Keywords: Neurofibroma; Nasal tip; Open rhinoplasty; Recurrence; Tumor

1. Introduction

Neurogenic tumors are known as any tumors that have origin from the neural tissue or from its covering. The two most common types are schwannomas and neurofibroma. Neurofibromas might be part of neurofibromatosis type 1 or sporadic. They also can be either typical or diffuse, with the diffuse variant being unusual variant with the skin and subcutaneous tissue being diffusely infiltrated.(1) Neurofibroma can arise in any anatomic location, however having a neurofibroma originating from the external nose more specifically the nasal tip is extremely rare and their management is considered challenging due to its position, the high rate of recurrence, the cosmetic challenge and the lack of ideal surgical approach due to the rarity of such condition.(2) We report a case of nasal tip neurofibroma in 11 years old female child.

2. Case Report

A 11 years old female child was referred to our otorhinolaryngology clinic from the dermatology clinic regarding a lesion over the nasal tip which is not responding to their medical management over a period of one month. History was taken from her guardians and they reported that the lesion over her nasal tip has been noticed few months ago and its slowly increasing in size without other complains such as pain or discharge from it and no history of nasal trauma related to it. The child is medically free, surgically free regarding the nose and she got no related nasal symptoms.

On examination of the nose, a soft rounded swelling was present over the right nasal tip measuring about (1.0*0.5) cm, the lesion was not tender, not mobile, firm in consistency and without skin changes or discoloration. Endoscopic examination of the nose was done and it was normal.

After discussion with her guardians the decision was made to excise the lesion using open rhinoplasty approach. Operation was done in January 2024 via an inverted V columellar incision and de-gloving over the nasal tip was done,

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then the soft tissue mass was removed by dissecting it from the underlying cartilage and from the overlying skin and skin was closed with nylon 5-0 stitches.

The excised lesion was sent for histopathology examination and their report confirmed that the lesion is neurofibroma, with immunohistochemistry results that S100 shows patchy positivity, and negative for CK, SMA, CD34.

We have been following our patient for 9 months so far, and there are no signs of lesion recurrence and with good cosmetic results, and now we follow her up on monthly basis until a period of one year regarding the recurrence of the lesion.

3. Discussion

Usually when a nasal mass or lesion present the patients seek medical advice early, this is due its location regarding the face being easily noticed which results in unpleasant look especially in the selfie photos era. Our patient presented early in which the lesion didn't cause effect on the cartilaginous frame work of the nose.(3)

Nasal lesions always represent a challenge regarding their treatment this due to their sensitive location, cosmetic considerations and the high recurrence rate.(4)

In our case the lesion was located at the nasal tip which is a major aesthetic subunit of the nose and adding to that it is more challenging since the skin over the nasal tip is thicker, more adherent, contains more sebaceous glands and also less flexible in relation to the nose subunits.(5) In general, the management of such lesions depends on a group of factors which are; the size and site of the lesion, being benign or malignant, tendency for recurrence, the expert of the treating tram and keeping in mind the patient's preference.(6)

In our case the lesion was small, and present at the right side of nasal tip near the dome, and major concern for the child and her guardian was to remove it with the best cosmetic results, this led us to use the open rhinoplasty approach with minimal exposure limited to the tip. While operating the challenge was that there is no definitive demarcation line of the lesion, so we removed it until the lower lateral cartilage was reached.

Surprisingly the lesion histopathological result showed it to be neurofibroma, which was never expected, even a computed tomography was done prior to surgery which was not informative regarding the lesion, and magnetic resonance image was not ordered due to the small size of the lesion, which in general in case of neurofibroma show a high signal on T2 weighted images, however a solid diagnosis cannot be achieved based on imaging only.(7)

Referring to the characters of neurofibroma, the high recurrence rate is the main concern and specially in our case since we were not aggressive in removing the lesion. This was explained to the guardian of the patient and that it needs close are regular follow ups regarding the recurrence and if that happened what the options of treatment that may have and that rarely neurofibromas may show malignant transformation.(8)

In 2007, Rameh et al. reported a case of nasal tip solitary plexiform neurofibroma in which was managed by open rhinoplasty approach. In his case the patient was a teenager with great attention to preserve the cartilaginous frame work so it will not interfere the growth and development of the patient.(9)

While reviewing all the documented cases of nasal tip lesions, the primary modality of management was surgical excision. Other means for treatment of neurofibroma at other anatomical subunits is Mohs micrographic surgery, that is mainly used in cases of recurrence. Mohs micrographic surgery is considered a useful technique for dealing with nasal lesions that is difficult to treat due to cosmetic and functional challenges.(10,11).

4. Conclusion

The open rhinoplasty approach gives a great access for removing nasal tip lesions and with good cosmetic results. Rare lesions should be kept in mind as differential diagnosis and pre-operative radiographic imaging can help the diagnosis if they are applicable.

Neurofibroma at the nasal tip are most likely not completely excised due to being located at cosmetic sensitive area, so this can give a high recurrence rate in which that affect the future management plan.

Compliance with ethical standards

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Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of informed consent

Informed consent from the guardian was taken only for discussion of the case and not permitted any image consent.

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