# Transnasal endoscopic excision of an isolated neurofibroma of the nasal septum*

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SUMMARY *Neurofibroma may occur in any parts of myelinated nerves having Schwann cells. However, it is extremely rare in the nasal septum. We have had experience with an isolated neurofibroma of the nasal septum which was successfully removed by transnasal endoscopic excision, and describe the clinical, endoscopic surgical and pathological features.*

*Key words: neurofibroma, nasal septum, transnasal endoscopic excision*

## INTRODUCTION

Neurofibroma is a neurogenic tumour arising from Schwann cells or peripheral tissues of nerve sheaths. It is usually presented with Von Recklinghausen's disease rather than as a solitary tumour (Batsakis, 1979; Hillstrom et al., 1990; Annino et al., 1991). It is commonly found in the head and neck and the flexoral surfaces of the upper and lower extremities. In the head-and-neck region, it often originates from the vestibular nerve. However, neurofibroma arising from the nose and paranasal sinuses are rare, and especially those from the nasal septum are extremely rare (Batsakis, 1979; Hillstrom et al., 1990).

The treatment of choice for neurofibroma in the nose and paranasal sinuses is complete excision via a lateral rhinotomy approach or a mid-facial degloving approach (Batsakis, 1979; Price et al., 1988; Stevens and Kirkham, 1988; Annino et al., 1991). However, if neurofibroma is present as a small solitary tumour, it is curable by adequate primary excision (Batsakis, 1979). Recently, the technique of endoscopic nasal surgery has rapidly developed, and transansal endoscopic excision of the primary lesion is successful when neurofibroma is a solitary tumour.

## CASE REPORT

A 58-year-old female patient presented with a history of progressive bilateral nasal obstruction, persistent nasal discharge, post-nasal drip, headache and hyposmia over the past 1 month. She had no history of epistaxis, pain, abnormal skin pigmentation nor a family history of neural tumour.

Diagnostic endoscopy revealed a firm, huge mass completely filling the right nasal cavity. The mass had a smooth mucosal surface, was ovoid in shape, and had a pale colour. It originated from the postero-superior portion of the right nasal septum.

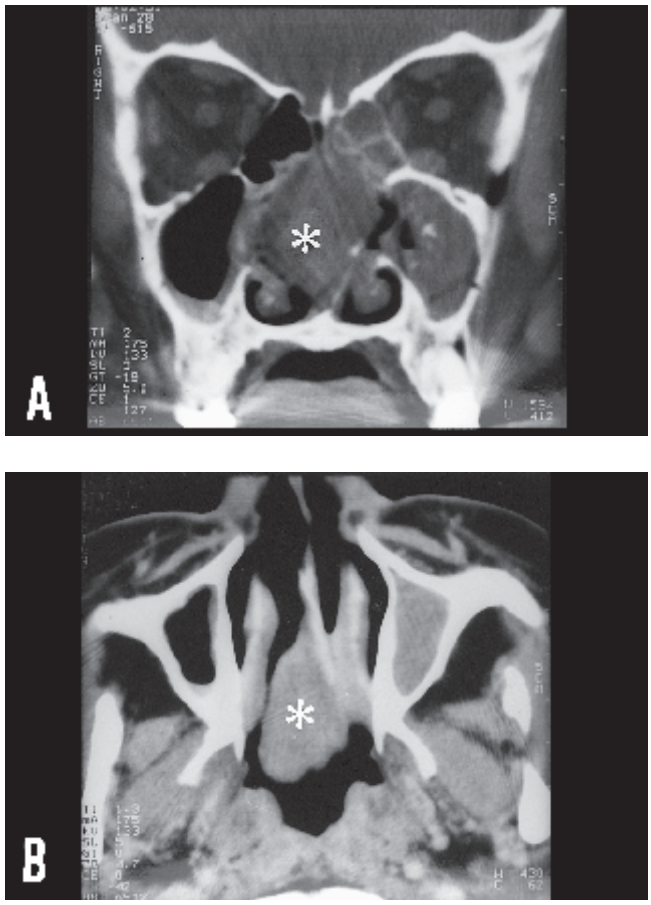


Figure 1. Contrast-enhanced paranasal sinus coronal (A) and axial (B) CT scans. A large soft-tissue density mass (*) is present in the right nasal cavity, right choanae and nasopharynx. The septum is deviated to the left side as result of a mass in the right nasal cavity. However, there is no evidence of bone destruction nor of extension of the lesion into the orbit or intracranial compartment.

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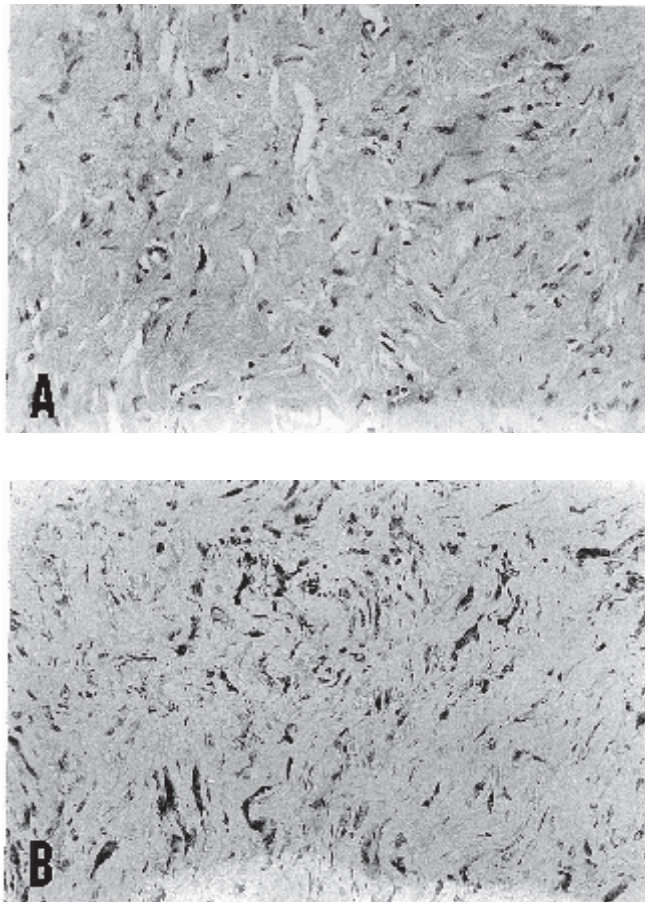


Figure 2. Histological findings of the tumour. A: Disorderly proliferation of collagen fibers and spindle-shaped tumour cells with wavy dark-staining nuclei (haematoxylin/eosin; ×400). B: The spindle-shaped tumour cells reveal immunoreactivity for the S-100 protein (×400).

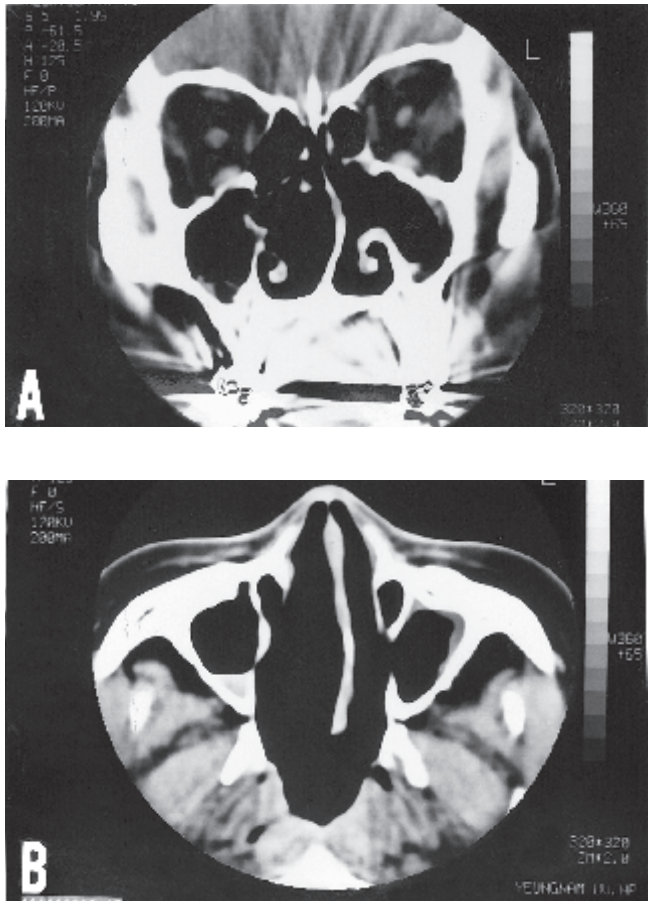


Figure 3. Post-operative contrast-enhanced paranasal sinus coronal (A) and axial (B) CT-scans, showing no recurrence of the pathologies after 12 months.

With nasopharyngoscopy, the mass almost filled both choanae. There was no regional lymphadenopathy nor evidence of ocular involvement.

Plain paranasal sinus X-rays showed an opacity in the right maxillary sinus and right nasal cavity without bony destruction. A CT-scan with contrast-enhancement demonstrated a large soft-tissue density mass in the right nasal cavity, right choanae and nasopharynx. The septum had a deviation to the left side as a result of the mass in the right nasal cavity (Figure 1). Also, soft-tissue density shadows were seen in the left ethmoid sinus, left frontal sinus and left maxillary sinus with punctuated calcification. However, there was no evidence of bony destruction or extension of the lesion into the orbit or intracranial compartment.

A punch biopsy was performed and this showed chronic inflammation. We thought that the abnormal mass in the nasal cavity was a benign tumour on the grounds of the pre-operative CT scan and diagnostic endoscopic findings. Consequently, the mass was removed by transnasal endoscopic surgery for definitive diagnosis and treatment. The mass arose from the postero-superior portion of the right nasal septum, occupied the right nasal cavity and extended to the nasopharynx and left nasal cavity. Endoscopic sinus surgery of the left paranasal sinuses was performed which revealed an aspergillosis.

The resected mass was not encapsulated but relatively circumscribed. The cut surface of the mass was pale, gray-white, solid and homogeneous. Histological examination of the mass showed combined proliferation of eosinophilic, thin, wavy collagen fibers lying in a disorderly pattern and spindle-shaped cells with a wavy, serpentine configuration and pointed ends. The stroma was infiltrated with lymphocytes and mast cells. Mitoses were not observed. The spindle-shaped tumour cells revealed immunoreactivity for S-100 protein (Figure 2).

The final pathological diagnosis was neurofibroma arising from the right nasal septum and aspergillosis in left maxillary sinus. After transnasal endoscopic surgical removal, no other surgical treatments have been performed. The post-operative course was uneventful. The aspergillosis of the left maxillary sinus was completely removed and no recurrence of neurofibroma has been observed during 12-month follow-up (Figure 3).

## DISCUSSION

Peripheral nerve-sheath tumours are divided into *neurofibroma*, *schwannoma* and *neurogenic sarcoma*. Neurofibroma and schwannoma are classified as benign, and neurogenic sarcoma as malignant (Batsakis, 1979; Hillstrom et al., 1990; Annino et al., 1991). All peripheral nerve-sheath tumours are believed to arise from Schwann cells. They often arise from sensory nerves and are found in the head and neck and the flexoral surface of the upper and lower extremities. However, they are rarely found in the nasal cavity and paranasal sinuses. In the nasal cavity and

paranasal sinuses, the great majority of the peripheral nerve-sheath tumours reported were schwannomas. A review of 430,000 pathological specimens by Perzin et al. (1982) revealed six neurofibroma involving the nasal cavity or paranasal sinuses. Almost all neurofibromas are associated with Von Recklinghausen's disease, so isolated neurofibromas without this association are extremely rare. Isolated peripheral nerve-sheath tumours occur commonly in females, in the fifth and sixth decades (Morris, 1989; Annino et al., 1991).

The neurofibroma occurring in the nose may be solitary or multiple. If neurofibroma are plexiform, they would be often part of neurofibromatosis type-I (Hellquist, 1990). 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 (Batsakis, 1979). A combined naso-ethmoid involvement is most common. It is followed, in order of frequency, by maxillary sinus, nasal cavity and sphenoid sinus (Robitaille et al., 1975; Batsakis, 1979). In the nasal septum, it is extremely rare, and nearly all have been solitary tumours.

Clinically, symptoms and signs are dependent on the site. Usually common symptoms and signs are nasal obstruction, epistaxis, facial pain, swelling, and proptosis. Schwannoma often produce a painful sensation and tenderness and are predominantly centrifugally distributed, whereas 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.

Histologically, the neurofibroma is non-encapsulated, poorly circumscribed with an ill-defined margin. This is typified by relatively hypocellular proliferation of bland, palely 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 (Perzin et al., 1982; Morris, 1989; Hillquist, 1990). In contrast, a schwannoma is a truly encapsulated tumour and shows two distinct patterns, Antoni-A and Antoni-B. The nuclei are arranged in pallisades with space between the rows, forming the so-called Verocay bodies.

The treatment of choice for benign peripheral nerve-sheath tumours is total local excision. Although neurofibroma is more invasive and more likely to recur after removal than schwannoma, recurrence is rare. Complete local excision is usually curative (Batsakis, 1979; Stevens et al., 1988), and prognosis is excellent, if the tumour is completely removed. Malignant transformation of neurofibroma is rare unless the patient has neurofibromatosis (Agarwal, 1979; Batsakis, 1979; Hellquist, 1990; McGee et al., 1992). In those situations malignant degeneration occurs in about 10% to 15% of the time (Devita et al., 1989; Annino et al., 1991).

## CONCLUSION

Neurofibroma is a peripheral nerve-sheath tumour. Solitary neurofibroma of the nasal septum is extremely rare, and curable by complete local excision via a transnasal endoscopic approach.

## REFERENCES

1. Agarwal MK (1979) Neurofibroma of the maxillary antrum. Oral Surg 48: 150-152.
2. Annino DJ, Domanowski GF, et al. (1991) A rare cause of nasal obstruction: A solitary neurofibroma. Otolaryngol Head Neck Surg 104: 484-488.
3. Batsakis JG (1979) Tumours of the Head and Neck. Clinical and Pathological Considerations. Williams Wilkins, Baltimore, pp. 313-333.
4. Devita VT, Hellman S, et al. (1989) Cancer. Principles and practice of oncology. In: Chang AE, Rosenberg SA, et al. (Eds.) Sarcomas of Soft Tissue. J.B. Lippincott Company, Philadelphia, pp. 1345-1398.
5. Hellquist HB (1990) Pathology of the Nose and Paranasal Sinuses. Butterworths, London, pp. 104-105.
6. Hillstrom RP, Zarbo RJ, et al. (1990) Nerve sheath tumours of the paranasal sinuses: Electron microscopy and histopathologic diagnosis. Otolaryngol Head Neck Surg 102: 257-263.
7. McGee JO, Isaacson PG, et al. (1992) Oxford Textbook of Pathology. Oxford University Press, Oxford, pp. 1892-1893.
8 Morris JH (1989) The nervous system. In: Cotran RS, Kumar V, et al. (Eds.) Robbin's Pathologic Basis of Disease. W.B. Saunders, Philadelphia, pp. 1445-1446.
9. Perzin K, Panyu H, et al. (1982) Nonepithelial tumour of the nasal cavity, paranasal sinuses and nasopharynx. Cancer 50: 2193-2202.
10. Price JC, Holliday MJ, et al. (1988) The versatile midface degloving approach. Laryngoscope 98: 291-295.
11. Robitaille Y, Seemayer TA, et al. (1975) Peripheral nerve tumours involving paranasal sinuses: A case report and review of the literature. Cancer 35: 1254-1258.
12. Stevens DJ, Kirkham N (1988) Neurofibroma of the paranasal sinuses. J Laryngol Otol 102: 256-259.

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