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Case Report
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# Nasal Neurofibroma with Cystic Degeneration: A Case Report

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## Abstract

**Introduction:** Neurofibroma originated from nasal cavity and paranasal sinuses are rare, and we report a rare case of nasal neurofibroma with cystic degeneration.

**Case Report:** A 61 years old female was found to have a large cyst in the left nasal cavity due to nasal obstruction. Eight months after cyst resection, she came to our hospital due to nasal and frontal distended pain, tumor found in the left nasal cavity. The tumor was resected completely under nasal endoscopy, and the postoperative pathology was neurofibroma. No recurrence was observed after one year follow-up.

**Discussion and Conclusion:** Nasal neurofibroma is rare and easily misdiagnosed, confirmed by immunohistochemistry. Owing to its characteristic of invasive growth, we should look for the tumor pedicle under the nasal endoscopy and remove the tumor completely, grinding the surrounding bone, and avoid recurrence.

**Keywords: Neurofibroma; Nasal cavity; Sinuses; Cystic degeneration**

## Introduction

Neurofibroma is a benign peripheral neuromatoid proliferative lesion originating from the peripheral nerve sheath, especially the endoneurium [1]. There was no significant difference between men and women, and the onset age was 20 to 40 years old. Tumors may exist in isolation but are usually closely associated with neurofibromatosis type I [2]. The primary neurofibroma in the nasal cavity is rare. We reported a rare case of neurofibroma with cystic degeneration in our department, and discussed its clinical manifestations, diagnosis, treatment and outcome.

## Case Presentation

A 61-year-old female patient was admitted to Nanjing Tongren Hospital for the first time due to nasal obstruction on January 23rd, 2018. A large cyst in the left nasal cavity was found by nasal endoscopy, CT and MRI examination (Figure 1). Therefore, left ethmoid sinus cyst resection and open sinus were performed under nasal endoscopy. One month after the operation, there was swelling and pain in the nasal and frontal. After several symptomatic treatments, there was no improvement. On September 17th, 2018, the patient was admitted to Tongren Hospital again. A neoplasm was found in the left frontal sinus under the outpatient nasal endoscopy, which was easy to bleed. A neoplasm biopsy was performed. The pathological results showed that spindle cell tumor. Combined with immunohistochemistry, neurofibroma and invasive growth were considered. The patient was admitted to our hospital for further treatment on October 15th, 2018. ENT examination showed that there was no deformity in the external nose, red neoplasm in the left nasal cavity, easy to bleed, no swelling of the turbinate, slightly right deviation of the nasal septum, and no tenderness in the paranasal sinus area. Neurological examination showed no abnormal, she had neither skin pigmentation, nor family history of neurofibroma. CT scan showed that soft tissue mass shadow in the left frontal sinus and ethmoid sinus was visible, and the adjacent bone resorption became thinner and the structure was irregular (Figure 2A). Enhanced MRI scan showed that the mass was significantly enhanced (Figure 2B). After completing other preoperative auxiliary examinations to eliminate surgical contraindications, on the second day after admission, the nasal neoplasm

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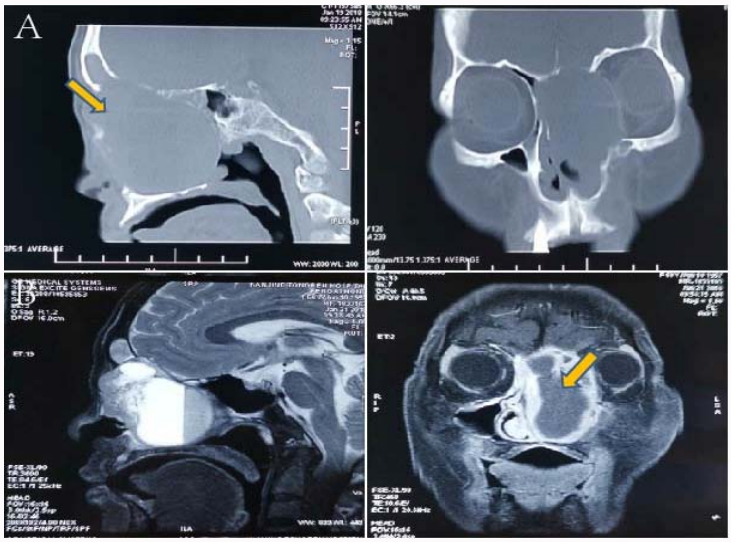


**Figure 1:** A: Large soft tissue shadow was seen in the left ethmoidal sinus (Yellow arrow), a cyst considered, compressing the surrounding bone and bone was defected on the ethmoidal top (CT). B: Large soft tissue shadow in the left ethmoidal sinus was visible (Yellow arrow), enhanced scan showed uneven enhancement (MRI).

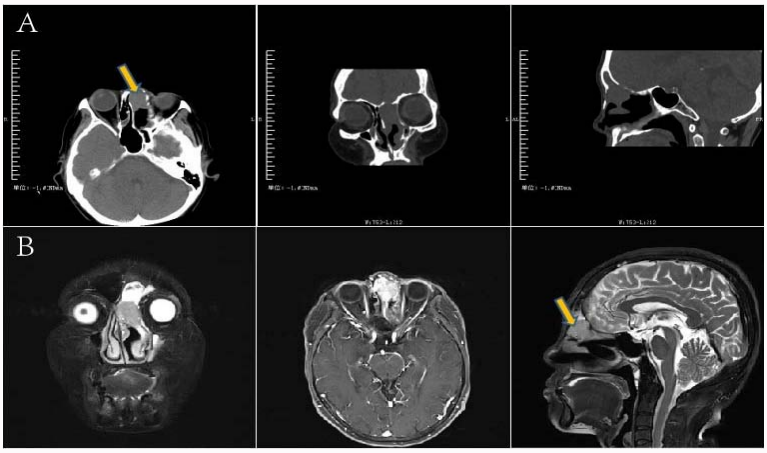


**Figure 2:** A: Soft tissue density shadow in the left frontal sinus and ethmoid sinus was visible (Yellow arrow), and the adjacent bone resorption became thinner and the structure was irregular (CT). B: After the operation of left ethmoid sinus, soft tissue mass shadow was observed in the left ethmoid sinus (Yellow arrow), with homogeneous signal, T1 equal signal, T2 slightly high signal, DWI high signal and ADC low signal. The mass was significantly enhanced after enhanced scanning (MRI).

resection and open sinus were performed under general anesthesia and nasal endoscopy. During the operation, the left nasal cavity was filled with red neoplasm. Most of the tumors were removed by low temperature plasma knife, the frontal recess was exposed, continue to ablate the tumor along the edge of the tumor. The tumor pedicle was located in the cribriform roof and extended to the nasal septum and cribriform plate. The tumors were completely removed, exposing the bone of the nasal septum, the bone of the skull base and the orbital cardboard. There was a skull base bone defect about 0.7 cm × 1.0 cm at the cribriform plate, and no obvious cerebrospinal fluid outflow. Mucosa edema and purulent secretions were observed in the left maxillary sinus, which were irrigated repeatedly with normal saline, and intraoperative bleeding was 50 ml. MRI was reexamined one week after operation showed that the tumor has been removed completely (Figure 3A). Nasal endoscopy was reviewed two weeks after operation showed that nasal discharge and mucosa edema (Figure 3B). Postoperative pathological results revealed that spindle cell proliferation showed positive reaction with S-100 and Ki-67 protein (Figure 4). After one year follow-up, there was no recurrence (Figure 3C).

## Discussion

Neurofibroma is one of the most common benign tumors of peripheral nerve, which first reported by Verocay. It is now believed that neurofibroma originate from nerve Schwann cells. Neurofibroma is common in young and middle-aged people, 90% of them are unilateral onset, and it can occur in any part of the body. In otolaryngology head and neck surgery, neurofibroma is more common in the throat and auditory nerve of the ear, however, the primary neurofibroma in the nasal cavity is rare, accounting for only 2.75% of the benign tumors in the nose. In the nasal cavity, the anterior part of the nose (nasal vestibule, middle nasal meatus) is common, most of neurofibroma come from inferior turbinate or nasal septum, it can involve the posterior nostril and pharynx [3,4]. In this case, the tumor originated from ethmoidal sinus. Nasal Neurofibroma often originates from maxillary branches and ophthalmic branches of the trigeminal

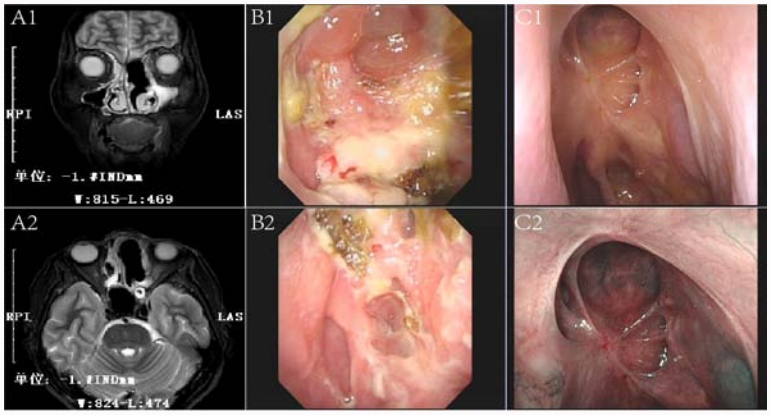


**Figure 3**: Postoperative reexamination. (A1-2) MRI was reexamined one week after operation showed that the tumor has been removed. (B1-2) Nasal endoscopy was reviewed two weeks after operation showed that nasal discharge and mucosa edema. (C1-2) Nasal endoscopy was reviewed one year after operation showed that nasal mucosa was epithelialized without tumor recurrence.

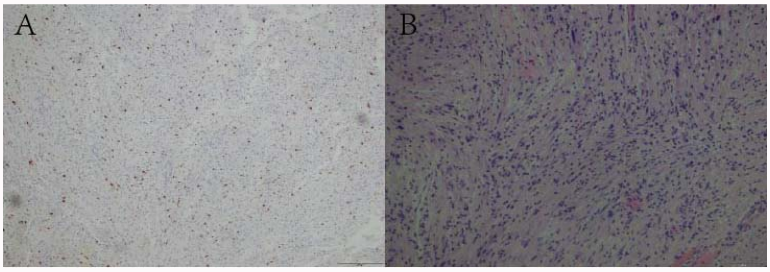


**Figure 4**: Postoperative pathological results. A: Microscopy examination showed that the tumor was well circumscribed and spindle cell proliferation (x200). B: Immunohistochemical study showed the spindle tumor cells revealed positive reaction with S-100 protein (x400).

nerve, the branches of the autonomic nerve [5]. Neurofibroma is mostly substantial, with different sizes and no complete capsule [6]. The tumor grows slowly, the course of disease is long, and the patients have no obvious symptoms in the early stage. Due to the different location of the tumor, the symptoms are nasal obstruction, epistaxis, headache, etc. In the later stage, the tumor increases and compresses the surrounding bone, making it thin and even destroyed; the tumor invades the peripheral structure along the bone destruction, such as paranasal sinus, orbit, pterygopalatine fossa and infratemporal fossa. On examination, there is gray or red mass in the nasal cavity, which is easy to be misdiagnosed as nasal polyp. The CT findings of paranasal sinuses shows a soft tissue density mass with CT value of 30 Hu to 50 Hu, which can be distinguished from general cyst, occasional calcification, adjacent tissues may show compression deformation and bone resorption. The CT manifestations of malignant neurofibroma have no significant difference with other nasal malignant tumors, so it needs to be combined with histopathological examination. Neurofibroma is mainly composed of Schwann cells and nerve fiber cells under microscopy. The tumor cells are spindle-shaped, and the intercellular space is filled with a large number of collagen fibers and mucoid substances, occasionally axons [1,7]. However, HE staining is often unable to determine the origin of tumor tissue, so the diagnosis still needs immunohistochemistry. The positive results of S-100 and Vimentin shows the tumor originate from nerve Schwann cells. SMA, desmin, CD34, CD117 negative exclusion of smooth muscle tumors, angiogenic tumors, stromal tumors, etc. In addition, neurofibroma must be distinguished from neurilemmoma and malignant peripheral neurilemmoma. Neurilemmoma has typical palisade cells, and the density of tumor cells is higher than neurofibroma. However, the cell morphology and nucleus of malignant peripheral neurilemmoma are pleomorphic. Our patient was initially diagnosed as a nasal cyst in Tongren Hospital, which it may be the blockage of the sinus orifice caused by tumor, which leads to inadequate drainage. As time goes on, a large cyst is formed. The cyst compresses the tumor to make it invisible, compresses the surrounding bone to make its absorption thin, and the imaging performance is not obvious. Finally, only the cyst is removed during the operation, and the scope of removal is not enough, not completely resection of residual tumor leads to recurrence. Neurofibroma is not sensitive to radiotherapy and should be completely removed by surgery as early as possible. According to the location, size and invasion range of the tumor, nasal endoscopic, lateral rhinotomy and lip-gingival groove approach can be used. Because of the advantages of nasal endoscopy such as clear field of vision, flexible perspective, simple operation and minimally invasive, Hirao reported that resection of neurofibroma of nasal cavity and paranasal sinuses under endoscopy is very effective [2]. With the development of imaging and endoscopic techniques, the understanding of the anatomy of the nasal cavity, paranasal sinuses and adjacent skull bases has been deepened; resection of neurofibroma of nasal cavity and paranasal sinuses under endoscopy is the preferred method. However, for the huge tumor, the boundary is often unclear, the capsule is incomplete, and the bone may be infiltrated, so it needs to be extended resection, and the tumor can be removed by the combination of nasal endoscopy and external nasal approach. Postoperative follow-up is very important for patients with neurofibroma. Femer reported that 2% to 5% of patients with neurofibromatosis type I turned into malignant peripheral neurilemmoma. It is necessary to resect completely as much as possible for malignant tumor, and adjuvant radiochemotherapy can improve the prognosis of patients [8]. In our case, there was no

recurrence after one year follow-up.

## Conclusion

The clinical symptoms of nasal neurofibroma are nonspecific and cannot be diagnosed by imaging examination alone. The diagnosis needs to be combined with pathological examination, especially there is a large cyst combined, the tumor may be concealed by cyst compression, and it is easy to be misdiagnosed as a nasal cyst according to CT and MRI findings. Therefore, careful exploration during the operation is equally important to ensure that there is no residue. Resecting the tumor completely is the key to prevent recurrence.

## References


1. Ferner RE, O'Doherty MJ. Neurofibroma and schwannoma. Curr Opin Neurol. 2002;15(6):679-84.
2. Hirao M, Gushiken T, Imokawa H. Solitary neurofibroma of the nasal cavity: resection with endoscopic surgery. J Laryngol Otol. 2001;115(12):1012-4.
3. Moreno PM, Meseguer DH. Solitary neurofibroma of the inferior nasal turbinate. Auris Nasus Larynx. 1998;25(3):329-31.
4. Berlucchi M, Piazza C, Blanzuoli L. Schwannoma of the nasal septum: A case report with review of the literature. Eur Arch Otorhinolaryngol. 2000;257(7):402-5.
5. Tall A, N'diaye, C, Diom ES. Solitary neurofibroma originating from the posterior nasal septum: Transnasal endoscopic resection. Eur Ann Otorhinolaryngol Head Neck Dis. 2015;132(4):223-5.
6. Sharma SB, Hong P. Solitary neurofibroma of the frontal sinus. Case Rep Otolaryngol. 2012;2012:373808.
7. Rameh C, Husseini S, Tawil A. Solitary plexiform neurofibroma of the nasal tip: Case report and review of the literature. Int J Pediatr Otorhinolaryngol Extra. 2007;2(2):116-9.
8. Ferner RE, Gutmann DH. International consensus statement on malignant peripheral nerve sheath tumors in neurofibromatosis. Cancer Res. 2002;62(5):1573-7.