

# Nasopharyngeal Carcinoma Mimicking Sinonasal Undifferentiated Carcinoma

## ABSTRACT

Nasopharyngeal carcinoma (NPC) is a tumor arising from the epithelial cells that covers the surface and lining of the nasopharynx. It is divided into keratinizing squamous cell carcinoma, and non-keratinizing carcinoma, which can be further divided into differentiated and undifferentiated subtypes. Symptoms of NPC can be obscure due to its anatomic location. On the other hand, sinonasal undifferentiated carcinoma defined as a highly aggressive and clinicopathologically distinctive carcinoma of uncertain histogenesis that typically presents with locally extensive disease. **The present report** is a case of 66-year-old Malay woman with underlying hypertension, hyperlipidemia and ischemic heart disease presented with swelling over the left upper palate for 5 months associated with the nasal symptoms. Cranial nerves **were** intact. Nasal endoscopy of the left nose revealed a mass occupying the left nasal cavity with histopathology of sinonasal undifferentiated carcinoma. However, CT scan result showed huge left nasopharyngeal carcinoma with local invasion and intracranial extension.

Keywords: nasopharyngeal carcinoma, sinonasal **carcinoma**

## INTRODUCTION

“Nasopharyngeal carcinoma (NPC) is a tumor arising from the epithelial cells that cover the surface and line of the nasopharynx” (1). “It is a distinct form of head and neck that differs from other malignancies of upper aerodigestive tract in **terms** of its etiology, epidemiology, pathology, clinical presentation, and response to treatment” (2). “There is an annual incidence of 0.5-2.0 cases in 100,000 in the US and Europe, although numbers as high as 25 per 100,000 may be seen in endemic countries such as China” (6). “In Malaysia, majority of the **cases** (75%) presented at stage III/IV, with neck mass being the most common presenting features (42%)” (3). “World Health Organization, divided NPC on a pathological basis into three histological subtypes i.e keratinizing squamous cell carcinoma, non-keratinizing carcinoma, which can be further divided into differentiated and undifferentiated subtypes” (4). “Symptomatically NPC most often presents with nasal and ear complaints from sinus congestion and epistaxis, to otalgia and tinnitus. Physical exam classically reveals a neck mass or masses, and cranial nerve involvement is not common” (5). “Treatment of early and locally advanced disease is typically nonsurgical because of the proximity of nearby vital structures. Thus, there is a strong role for non-invasive therapy in the form of radiation ± chemotherapy, which is considered standard of care for this disease” (6).

“Sinonasal undifferentiated carcinoma (SNUC) is a poorly differentiated carcinoma that arises from Schneiderian mucosa which lines the nasal cavity and paranasal sinuses. It is a very rare disease with unknown exact incidence rate. SNUC first described by Frierson and colleagues in 1986, is a high-grade, round cell neoplasm of the sinonasal tract” (7). “WHO classified SNUC as a highly aggressive and clinicopathologically distinctive carcinoma of uncertain histogenesis that typically presents with locally extensive disease” (8). “Usually patients present with symptoms such as nasal obstruction and epistaxis, and due to its invasive nature, it results in proptosis, cranial nerve palsies, visual disturbance, and pain” (9). The ideal therapy for SNUC remains controversial, but multimodality therapy, including neoadjuvant chemotherapy followed by surgery in resectable cases, may result in improved prognosis.

## CASE PRESENTATION

A 66 year old Malay woman with underlying hypertension, hyperlipidemia and ischemic heart disease was referred by OMFS team for expert opinion on left maxillary sinus as noted haziness over left maxillary sinus in CBCT maxilla and mandible. Patient initially presented with painless swelling over the left upper palate for 5 months associated with left eye watery discharge for 3 months. Patient also complained of frequent rhinorrhea of the left nose associated with nasal blockage and epistaxis for 1 year. Otherwise, there was no cacostmia or anosmia. Furthermore, she also complained of bilateral hearing loss with tinnitus bilaterally and more over the left ear.

On examination, there was visible facial asymmetry over the left cheek due to swelling with intact facial nerve as shown in figure 1. Other cranial nerves were also intact. Intraorally noted swelling at the left upper palate extending until the soft palate as shown in figure 2. There were no palpable neck lymph nodes and systemic examination was normal. Nasal endoscopy revealed polypoidal nasal mucosa with mass occupying left nasal cavity as shown in figure 3. There was contact bleeding of the mass upon stimulation. Bleeding was secured with ribbon gauze soaked with cocaine. Right nasal cavity was congested with hypertrophied inferior and middle turbinate. Biopsy was planned after patient withheld his glyprin for 5 days. Pure tone audiometry showed mild to moderate hearing loss with notch at 4-8kHz over right ear with tympanometry type C and moderate mixed hearing loss over the left ear with tympanometry type B.



**Fig. 1:** Image showing swelling over left cheek

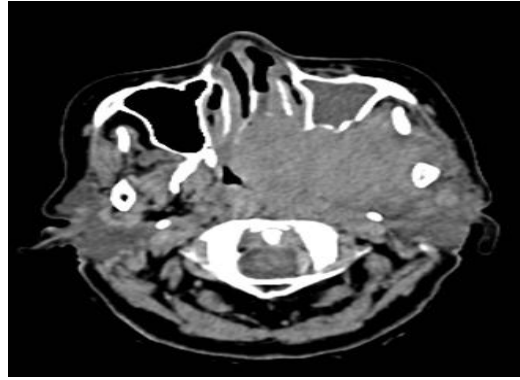


**Fig. 2:** Swelling over the left palate

Biopsy of the left nasal cavity's mass was taken on 5<sup>th</sup> of September 2018. Patient was admitted with BIPP inserted over the left nasal cavity after the procedure as noted oozing of blood over the biopsy site. However, patient was able to be discharged on the next day. Histopathology (HPE) result came out as sinonasal undifferentiated carcinoma. Patient was then treated as sinonasal undifferentiated carcinoma and was scheduled for CT base of skull till abdomen. However, CT scan reported as highly suggestive of a huge left nasopharyngeal carcinoma with local invasion and intracranial extension. Case was discussed back with pathologist as HPE result was contradicting with radiological findings and was told to correlate with clinical and radiologically as it still can be NPC.



**Fig. 3: Mass over left nasal cavity**



**Fig. 4: CT scan revealed mass originate from left nasopharynx**

This patient was also seen by ophthalmology team and treated as left nasolacrimal duct obstruction most probably secondary to compression of left nasal mass.

Patient was then referred to oncology team as nasopharyngeal carcinoma stage T4N2M0 for chemoradiotherapy.

## DISCUSSION

“The nasal cavity and paranasal sinuses are referred to as the sinonasal tract, which is anatomically and embryologically different from the nasopharynx. They form a functional unit, which is lined by stratified squamous, respiratory-type pseudostratified columnar, and transitional (intermediate) epithelium” (10).

NPC presents most commonly as neck lump (60%), ear fullness (41%), hearing loss (37%), epistaxis (30%), nasal obstruction (29%), headache (16%), ear pain (14%), neck pain (13%), weight loss (10%), diplopia (8%). Sore throat and throat pain is rare presentation and its incidence is not documented(11).

“The most common initial symptoms of SNUC are epistaxis, facial pain, and nasal obstruction. More than one-half of all patient exhibit symptoms related to the eye, e.g., proptosis, cranial nerve palsies, diplopia, or decreased visual acuity. Because many of the early symptoms are similar to those of benign sinus disease, patients often delay seeking treatment and ultimately present with advance-stage disease” (12).

The present case revealed that, patient main complaint was swelling over the left palate which is quite unusual presentation for NPC. However, patient also had complained of left nasal blockage, epistaxis and left ear fullness and reduces hearing which can be both presentation of NPC and SNUC. Patient denied having any eye symptoms except for left eye discharge.

Diagnosis of NPC is made by the help of imaging studies and histopathology in addition to nasal endoscopy. Further in this case, nasal endoscopy had limitation in view the mass obstructing the view made difficulty in identifying the origin of the mass. The initial histopathological result from the mass reported as sinonasal undifferentiated carcinoma. The diagnosis of NPC was confirm after CT scan showed mass arising from left side of nasopharynx with involvement of parapharyngeal space and obliterating the left fossa of rosenmuller. The mass also erodes left wall of maxillary sinus with obliteration of lateral and medial pterygoid plate.

"The most challenging differential diagnosis for SNUC include undifferentiated NPC. The cytologic features of undifferentiated NPC is similar to SNUC which are characterized by the presence of single cells, small loose clusters and occasional larger clusters, variably prominent nucleoli, and scant neoplasm. The clinical facts should aid in the differential diagnosis. The separation of these lesions is important because the mainstay of treatment for NPC is radiation therapy (often alone); the prognosis of NPC is much better than that of SNUC, 5-year-survivals around 60%" (7).

## CONCLUSION

To reach presumptive diagnosis, the presenting features and symptomatology and imaging studies with the help of histopathological examination is important to get a correct diagnosis and timely intervention. Correct diagnosis will direct the clinician towards proper management or proper outcome.

## Ethical Approval:

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

## CONSENT

Informed consent was taken from the patient that involve in this study.

## REFERENCES

1. Gupta R, Mohindroo NK, Azad R. Nasopharyngeal Carcinoma with uncommon Presentation. *Int J Head Neck Surg* 2017;8(1):34-36
2. Keiji Tabuchi, Masahiro Nakayama, Bungo Nishimura, et al. Early Detection of Nasopharyngeal Carcinoma. *International Journal of Otolaryngology* Volume 2011, Article ID 638058, 6 pages
3. Mohamad I, Kosha MY. Submandibular mass a rare presentation of advanced nasopharyngeal carcinoma. *Malays Fam Physician* 2013;8(3):40-2
4. Sandhu KS et al. *Int J Otorhinolaryngol Head Neck Surg*. 2017 Oct;3(4):1117-1119
5. Baba M, Dominguez LW, Patel A (2016) Atypical Presentation of Nasopharyngeal Carcinoma. *Hematol Transfus Int J* 2(4): 00041
6. Courtney Pollard III, Shane M. Mesko, Lawrence E. Ginsberg, et al. Nasopharyngeal carcinoma with intranasal spread. *Clinical Case Reports* 2017; 5(10):1682-1688
7. Andrew M. Bellizzi, MD, T. David Bourne, et al. The cytology features of Sinonasal undifferentiated Carcinoma and Olfactory Neuroblastoma. *Am J Clin Pathol* 2008;129:367-376
8. Bruce M. Wenig, MD. Undifferentiated Malignant Neoplasms of the Sinonasal Tract. *Arch Pathol Lab Med* Vol 133, May 2009
9. Pradeep N, Ghorpade R. Sinonasal undifferentiated carcinoma with intracranial extension: Case report. *J Sci Soc* 2016;43:41-3
10. Bhattacharya J., Goswami BK et al. Clinicopathological study, mass, sinonasal tract, nasopharynx. *The Egyptian Journal of Otolaryngology* 2015, 31:98-104
11. *Indian J. Otolaryngol. Head Neck Surgery*. (January-March 2018) 60,82-83

12. Judith Gorelick, M.D., Donald Ross, M.D., et al. Sinonasal Undifferentiated Carcinoma: Case series and Review of the literature. *Neurosurgery*, Vol. 47, No. 3, September 2000