International Journal of Otorhinolaryngology and Head and Neck Surgery
*Purushotman R et al. Int J Otorhinolaryngol Head Neck Surg. 2021 Nov;7(11):1821-1824*
http://www.ijorl.com
pISSN 2454-5929 | eISSN 2454-5937

DOI: https://dx.doi.org/10.18203/issn.2454-5929.ijohns20214237

**Case Report**

# Nasopharyngeal mucoepidermoid carcinoma: a case report and review of its current management

**Purushotman Ramasamy*, Vigneswaran Kumarasamy, Pathma Letchumanan, Harvinder Singh Dhalip Singh**

Department of Otolaryngology and Head and Neck Surgery, Hospital Raja Permaisuri Bainun, Ipoh, Malaysia

**Received:** 10 May 2021
**Revised:** 09 October 2021
**Accepted:** 12 October 2021

***Correspondence:**
Purushotman Ramasamy,
E-mail: purushotman85@gmail.com



## ABSTRACT

Salivary gland tumours arising at the nasopharynx is highly infrequent. Among them the commonest is mucoepidermoid carcinoma (MEC). Reports with a larger number of patients are often from Asian countries where nasopharyngeal squamous cell carcinoma is also predominant. Although nasopharyngeal MEC (NMEC) is a disease of adults, sporadic cases amongst children have been reported. We report a case of a 32 years old man presented with complaining of intermittent epistaxis over a year. His nasal endoscopy showed friable polypoid tumour at the right choanae. Histopathology revealed a NMEC and he underwent endoscopic endonasal nasopharyngectomy with adjuvant radiotherapy. The optimal treatment for NMEC is arbitrary due to the lack of evidence. However, unlike most sinonasal malignancies, NMEC has the tendency to manifest itself early and has good response to the treatment. Therefore, in this article we describe the clinical features and justifications for the selection of treatment options including surgical and non-surgical therapies and including the role of neck dissection.

**Keywords:** Nasopharynx, Mucoepidermoid carcinoma, Nasopharyngectomy

## INTRODUCTION

MEC is a well-understood tumour in the head and neck region. The salient histopathological features of MEC were first described by Stewart et al in 1945.[1] MEC occurring other than in the salivary gland are reported to have lung, nasal cavity and paranasal sinuses. NMEC is reported in less than 20 literatures. Among 2% of the salivary gland malignancy arising at the nasopharynx, MEC accounts for 15% of them.[2] Its rarity caused difficulty in demonstrating the gender predilection as there were disputes between different sources.[3,4] It commonly occured between the second to sixth decade of life. Our patient presented in his third decade with significant epistaxis over a period of one year before he was seen in an otolaryngology unit.

## CASE REPORT

A 32 years old man suffered recurrent, intermittent bilateral epistaxis for a year. At each episode, he suffered bleeding of about 100 cc and stopped only after seeking help in a nearby clinic. He had progressively worsening nasal block and accompanying hyposmia apart from right ear fullness. He had no neck swelling, no constitutional symptoms. He denied trauma, not on any drugs and did not smoke.

On examination, he was alert and his haemodynamics were stable, no neck swelling. A rigid nasal endoscope showed a friable, exophytic, polypoidal fleshy mass from the right nasopharynx obstructing the right choanae completely (Figure 1 A and B). Right fossa of rossenmuller (FOR) and eustachian tubes (ET) were obliterated by the tumour. The tumour crossed the

midline and extended to the left nasopharynx. However, the left FOR and ET were patent. The right tympanic membrane appeared retracted. However, the left ear, oral cavity, throat were normal on examination.

Office biopsy of the mass showed a low-grade mucoepidermoid carcinoma of the nasopharynx. Magnetic resonance imaging revealed a hyperintense lesion measuring 3.6×4.0×3.9 cm with necrotic centres and bleeding within. The tumour was seen occupying the bilateral fossa of rossenmuller (Figure 2). Multiple subcentimeter neck nodes were present and the largest was at the contralateral neck, measuring 1.3×1.5 cm. No bone erosion was seen.

The patient underwent an endoscopic nasopharyngectomy to remove the tumour. However, the tumour tissue was tough to be removed with microdebrider or cold steel, particularly at the posterior septum. The tumour also showed no clear delineation to the bony septum. The post-operative specimen reported no perineural or lymphoreticular invasion, but as we had expected, the tumour cells were positive at the edges of the tumour.

As further surgery was deemed ineffective, he subsequently underwent radiotherapy at the nasopharynx at a dose of 66 gy/33 #m for 6.5 weeks. The neck was spared as the tumour was low grade. The patient was disease and recurrence-free for 18 months (Figure 1C).

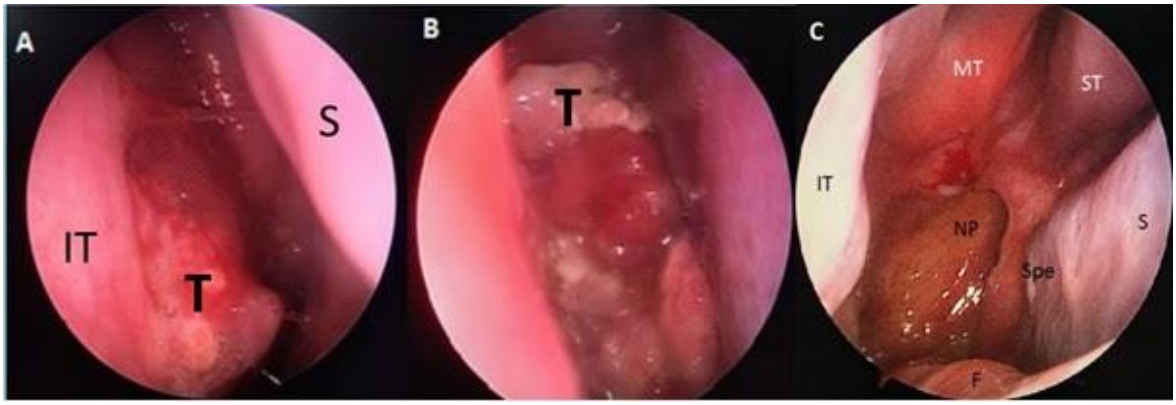


**Figure 1: Right nasal endoscope (A) tumour (T) in the anterior nasal cavity; (B) tumour obliterating the chonae posteriorly; (C) post op 18 months; IT: inferior turbinate, S: septum, MT: middle turbinate, ST: superior turbinate, Spe: septum-posterior end where septectomy done, NP: nasopharynx, F: floor of the nose.**

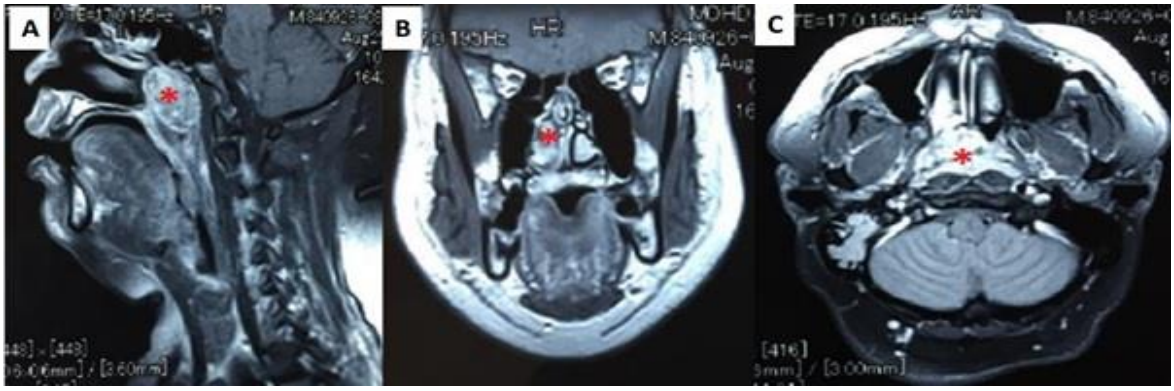


**Figure 2: MRI paranasal sinuses T1 image * showing the tumour (A) sagittal view: mixed signal showing intratumour bleeding; (B) coronal view: posterior cut showing fully obstructed choanae; (C) axial view: involving the bilateral for; no clear plane seen between the tumour and right tensor palatine, right medial pterygoids and bilateral levator palatine muscles.**

## DISCUSSION

### *Clinical background*

NMEC has the advantage of manifesting itself earlier than the nasopharyngeal squamous cell carcinoma.[3] Commonest symptoms were epistaxis, nasal block, hyposmia and hearing loss. It is also was not uncommon to be presented with neck swelling. Such presentation in a patient should immediately prompt a clinician for necessary investigations to enable early detection of the pathology.

Macroscopically it was challenging to differentiate it from the other type of nasopharyngeal malignancies. However, it tend to appear as pink-greyish, polypoidal with areas of necrosis. The breaching of the overlying mucosa may or may not be present. Intratumor haemorrhage and necrosis might be present.

Histology showed the typical appearance of mucin and glycogen containing epidermoid cells within the cytoplasm; arranged closely. The glycogen containing cells will appear as clear cells. Mucin staining (MUC1, p63, cytokeratins) and glycogen staining (periodic acid-schiff) will be positive.[2] MEC was graded according to the solid and cystic components, necrotic areas, mitotic figures and perineural invasion.

Computed tomography will help identify bony involvement, although this was rare. Magnetic resonance imaging (MRI) will usually demonstrate hyperintense irregular lesions on T1 weighted image. MRI was also beneficial in differentiating the actual tumour volume from the retention of secretion in the involved sinuses, and to know the involvement of the periorbita, cavernous sinuses and the involvement of the dura.

***Management***

Treatment of NMEC is cumbersome due to several reasons. To date, there is no common consensus in treating NMEC. The rarity of this condition caused an extreme lack of experience and therefore the data to arrive at one. The readily available literatures were not convincing in numbers to make conclusions. Furthermore, they tend to lump all salivary gland malignancies together. Thus, it might not reflect the actual outcome of treating NMEC.

Currently, various treatment options were adopted according to the locally available expertise and facilities. As most NMEC tend to present early, this gave us the best chance to tackle it at an early stage. Thus, correct treatment was crucial to deliver the best possible disease or recurrence-free life to the patients.

***Primary surgery and elective neck dissection***

The complex anatomy of the nasopharynx at the skull base caused primary surgical excision of the tumour almost not widely accepted in the past. The tumour, which was often large, posed a technical challenge in the surgical approach and obtaining a clear post excision margin. This was often due to the diffuse irregular lesions involving the skull base, posterior pharyngeal wall, posterior septum, paranasal sinuses and orbit.

The post-operative complications such as failure of flap uptake, epistaxis and velopharyngeal insufficiency were challenging to manage. Open surgeries to reach nasopharynx such as lateral infratemporal fossa approach, maxillary swing and transoral approaches were often cosmetically mutilating.

However, with the recent advancement in endoscopy and haemostatic agents, primary excision was no longer a nightmare for both the surgeon and the patient. We advocate that primary nasopharyngectomy should be performed in all patients presenting at early-stage disease. Although reaching cancer-free margins was almost impossible, the literature showed promising outcomes in expert hands when the patients were given post-operative radiotherapy.[3]

Elective neck dissection was not recommended in patients with N0 salivary gland malignancy. However, the occult neck disease was shown to be as high as 47% in patients with nasopharyngeal salivary gland malignancy.[5] Due to the high occult incidence, authors preferred that it was wise to perform a single seating elective supraomohyoid neck dissections in N0 patients. However, we did not perform this in our patient as the patient has refused to consent.

***Radiotherapy***

The effect of radiotherapy on NMEC was not convincing in available literatures. Traditionally MEC was believed to be radioresistant irrespective of the severity. However, recent evidence had shown that radiotherapy gave reasonable locoregional control after the primary surgery.[3,5,6] MEC on the other region such as larynx was even shown to achieve remission with radiotherapy alone.[3] The advent of intensity-modulated radiotherapy reduced the adverse effects resulting from conventional radiotherapy.[7] The role of brachytherapy had not been widely discussed. We recommend radiotherapy to be utilised as adjuvant therapy in patients undergoing primary excision or as a primary modality when the tumour was inoperable.

***Genetic discovery***

MAML2 gene was shown to be present in all MEC.[8] However, the presence of it was not correlated to the grade of malignancy. Further studies need to be done to identify possible treatment options. MUC1 and MUC4 expression identified in MEC correlated to its grades; high grade and low grade, respectively.[9,10] These were beneficial in predicting the recurrence and disease-free survival rates. To date, NMEC had no EBV association. Although the distant metastases are up to 25%, the prognosis seems promising, 90% in early-stage disease.[3,6]

## CONCLUSION

The authors find that the currently available data is extremely shortage of volumes to arrive at any conclusion about treatment. Given the good outcome post-treatment in NMEC, we advocate creating a regional database specific to NMEC. Regional collaboration is vital,

especially in Asia, where the incidence of nasopharyngeal malignancy is also higher.

## ACKNOWLEDGEMENT

We would like to thank the Director General of Health, Malaysia, for permission to publish this paper.

*Funding: No funding sources*
*Conflict of interest: None declared*
*Ethical approval: Not required*

## REFERENCES

1. Stewart FW, Foote FW, Becker WF. Muco-epidermoid tumors of salivary glands. Ann Surg. 1945;122(5):820-44.
2. Kusafuka K, Takizawa Y, Iida Y, Ebihara M, Onitsuka T, Kameya T. Primary nasopharyngeal mucoepidermoid carcinoma in Japanese patients: two case reports with histochemical and immunohistochemical analysis and a review of the literature. Virchows Arch. 2007;450(3):343-8.
3. Zhang XM, Cao JZ, Luo JW, Xu GZ, Gao L, Liu SY, et al. Nasopharyngeal mucoepidermoid carcinoma: a review of 13 cases. Oral Oncol. 2010;46(8):618-21.
4. Hemalatha AL, Kumar HKS. Nasopharyngeal mucoepidermoid carcinoma - a common entity at an uncommon location. J Clin Diagn Res. 2014;8(1):164-5.
5. Schramm VL, Imola MJ. Management of nasopharyngeal salivary gland malignancy. Laryngoscope. 2001;111(9):1533-44.
6. Ollero JM, Morón AH, Luis AM, Sánchez SM, Nazarewsky AA, López MJ, et al. Nasopharyngeal mucoepidermoid carcinoma: a case report and review of literature. Rep Pract Oncol Radiother. 2012;18(2):117-20.
7. Rosenbluth BD, Chou WW, Lee NY. IMRT for carcinomas of the nasopharynx. In: Bortfeld T, Schmidt-Ullrich R, DeNeve W, Wazer DE, eds. Image-guided IMRT. Berlin, Heidelberg: Springer; 2006.
8. Saade RE, Bell D, Garcia J, Roberts D, Weber R. Role of CRTC1/MAML2 translocation in the prognosis and clinical outcomes of mucoepidermoid carcinoma. JAMA Otolaryngol Head Neck Surg. 2016;142(3):234-40.
9. Seethala RR, Dacic S, Cieply K, Kelly LM, Nikiforova MN. A reappraisal of the MECT1/MAML2 translocation in salivary mucoepidermoid carcinomas. Am J Surg Pathol. 2010;34(8):1106-21.
10. Behboudi A, Enlund F, Winnes M, Andrén Y, Nordkvist A, Leivo I, et al. Molecular classification of mucoepidermoid carcinomas-prognostic significance of the MECT1-MAML2 fusion oncogene. Genes Chromosomes Cancer. 2006;45(5):470-81.

**Cite this article as:** Purushotman R, Vigneswaran K, Pathma L, Harvinder S DS. Nasopharyngeal mucoepidermoid carcinoma: a case report and review of its current management. Int J Otorhinolaryngol Head Neck Surg 2021;7:1821-4.