407

# CASE OF SINONASAL NEUROENDOCRINE TUMOUR GRADE 2 WITH SELLAR EXTENSION MANAGED WITH ENDOSCOPIC ENDONASAL RESECTION AND SKULL BASE RECONSTRUCTION

Lee King Peng, Ng Wei Ping, Nik Mohd Yunus Mohammad, Elang Kumaran Krishnan, Azmi Alias

HOSPITAL KUALA LUMPUR

ENDOKL2024

## INTRODUCTION

Sinonasal neuroendocrine tumor (SNET) is a rare neoplasm with an annual incidence of 0.4 cases per 100000 individuals. The tumours can exhibit varying degrees of extensions, invasions and systemic involvement.

## CASE REPORT

A 42-year-old male presented with nasal obstruction. Nasoendoscope revealed intranasal mass. MRI revealed large expansile mass occupying the sphenoid sinus with extension to nasopharynx, sella turcica, bilateral cavernous sinus, posterior choanae and clivus. PET/CT study revealed no distant metastasis.

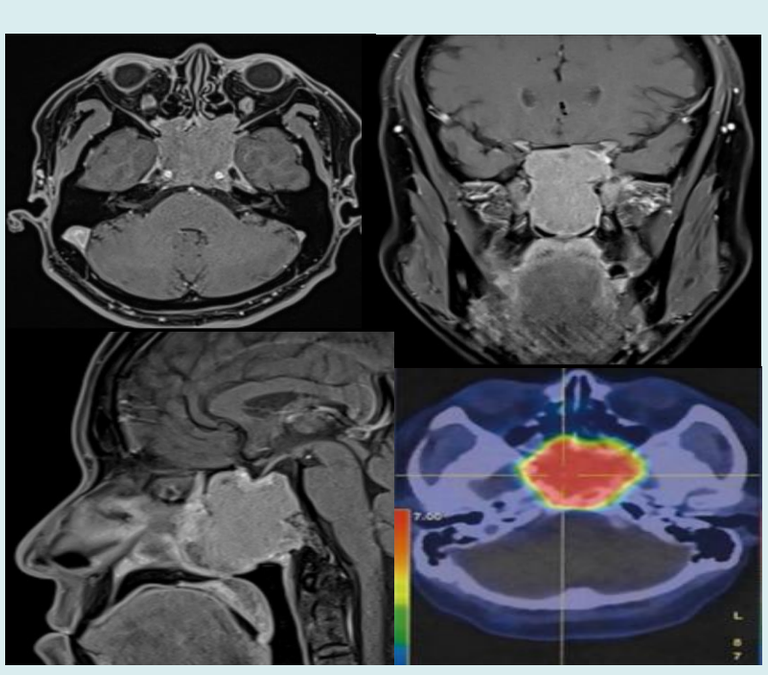

**Figure 1.** MRI and PET/CT findings

Endonasal biopsy results were suspicious of neuroendocrine tumour due to positivity for Synaptophysin and Chromogranin A. Subsequently, he underwent endoscopic endonasal resection of tumour and skull base reconstruction with lateral nasal flap. Intraoperatively, there was reddish soft vascular tumour arising from posterior nasal septum extending to bilateral nasal cavity, posterior choanae, sphenoid sinus and sellar cavity. Gross total resection was achieved. Postoperatively, patient was extubated with no deficits or Cerebrospinal fluid (CSF) leak.

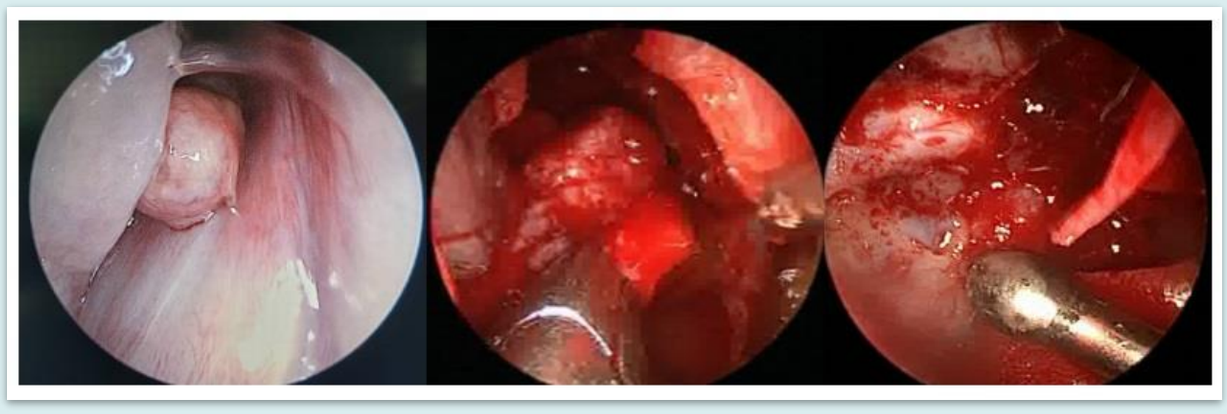

**Figure 2.** Intraoperative findings.

Histopathological analysis revealed neuroendocrine atypical carcinoid tumour (Grade II). Ki67 index 5-10%.

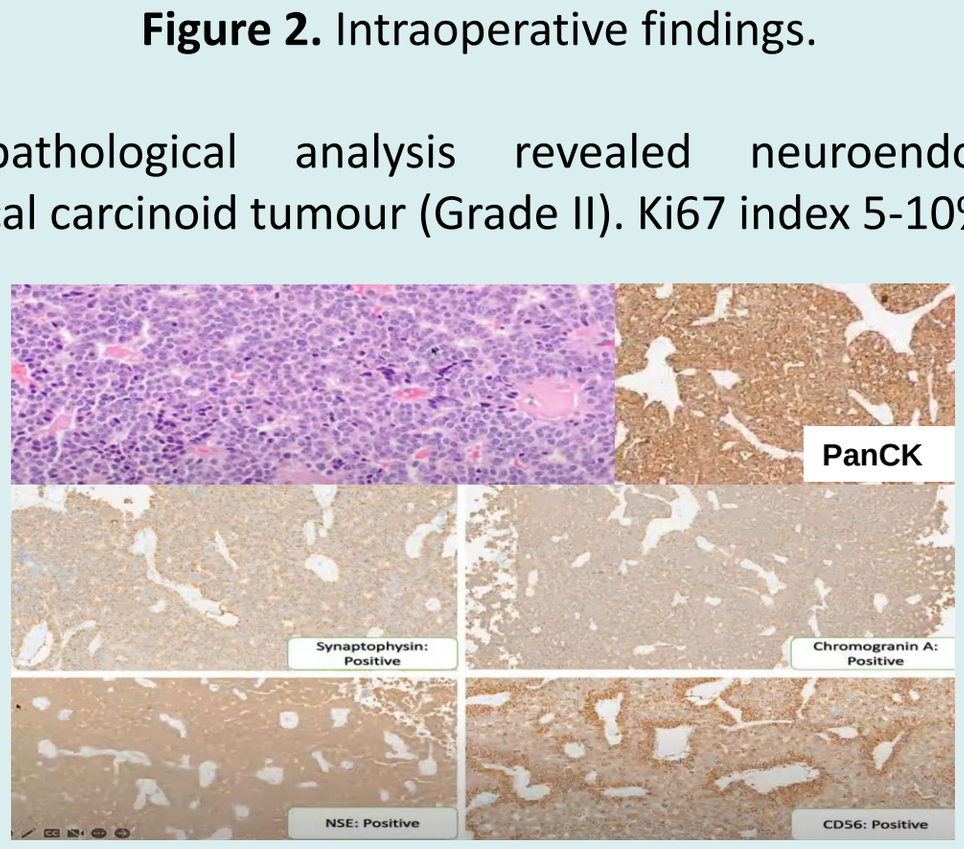


**Figure 3.** Histopathology and Immunohistochemistry

Patient then underwent 30 cycles of radiotherapy. He remained under surveillance, and as of his current 2-year follow-up, there is a small residual lesion in the right intrasellar region. The Growth Hormone and Serum Chromogranin A levels have remained stable.

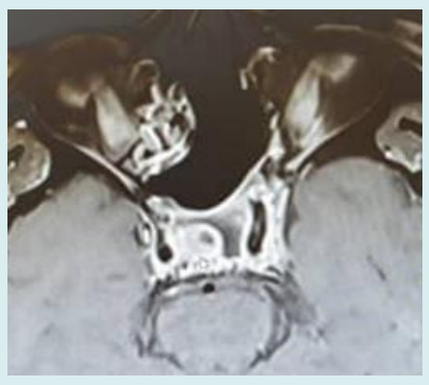

**Figure 4.** Latest MRI showed stable small right intrasellar residual lesion.

## DISCUSSION

SNET can occur at any age but are more commonly diagnosed in adults with slight male predominance. atients often present with symptoms such as nasal obstruction, epistaxis (nosebleeds), facial pain or pressure, and changes in vision or smell. It can be classified into well, moderately or poorly differentiated forms with further subcategorizations. Survival rates for the tumor can vary widely depending on the tumor subtype and stage at diagnosis.. Overall, SNET tend to have a relatively poor prognosis compared to some other head and neck malignancies due to their aggressive nature and propensity for local recurrence and distant metastasis. For the poorly differentiated form, 5-year survival rate is 35.9%. Treatment usually involves a multidisciplinary approach, including surgery, radiation therapy, and sometimes chemotherapy. The choice of treatment depends on factors such as tumor size, location, histological subtype, and extent of disease.Due to its rarity, there is no clear management guideline. Due to the rarity of SNETs, ongoing research efforts are focused on improving our understanding of the disease, identifying prognostic factors, and developing targeted therapies to improve outcomes for patients with these tumors.

## CONCLUSION

The diagnosis of SNET, though uncommon, should always be considered, especially in adolescents presenting with a Sinonasal mass. Maintaining a high index of suspicion for this rare pathology and performing a complete resection followed by adjuvant oncology treatment is the key to good outcome.

## REFERENCES

1.Bell D. Sinonasal Neuroendocrine Neoplasms: Current Challenges and Advances in Diagnosis and Treatment, with a Focus on Olfactory Neuroblastoma. Head Neck Pathol. 2018 Mar;12(1):22-30.

2.Wani SQ, Dar IA, Khan T, Lone MM. Primary Sino-nasal Neuroendocrine Carcinoma: A Rare Tumor. Cureus. 2019 Feb 27;11(2):e4144.

3.van der Laan TP, Iepsma R, Witjes MJ, van der Laan BF, Plaat BE, Halmos GB. Meta-analysis of 701 published cases of sinonasal neuroendocrine carcinoma: The importance of differentiation grade in determining treatment strategy. Oral Oncol. 2016 Dec;63:1-9.