

# Extensive HPV-Related Multiphenotypic Sinonasal Carcinoma: a Clinico-Radiological Case-Report with Full Endoscopic Surgical Outcome

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## Introduction

- HPV-related multiphenotypic sinonasal carcinoma (HMSC) is a relatively new diagnosis with fewer than 100 cases reported in the English literature [1-3].
- Potential mimickers include adenoid cystic (AdCC) and basaloid squamous cell carcinomas, which makes immunohistochemistry and HPV DNA identification critical to differentiate between these diagnoses [4].
- The slow and indolent evolution of HMSC, characterized by adjacent **structure remodeling**, is important in treatment planning [1, 5].
- Few case reports and case series have described the radiological and surgical correlations of HMSC tumors.

## Objectives

- To describe the case of the largest HMSC tumor reported in the English literature.
- To describe the radiological and surgical correlations in a large tumor that was successfully managed with an endoscopic surgical resection despite significant radiological extension.

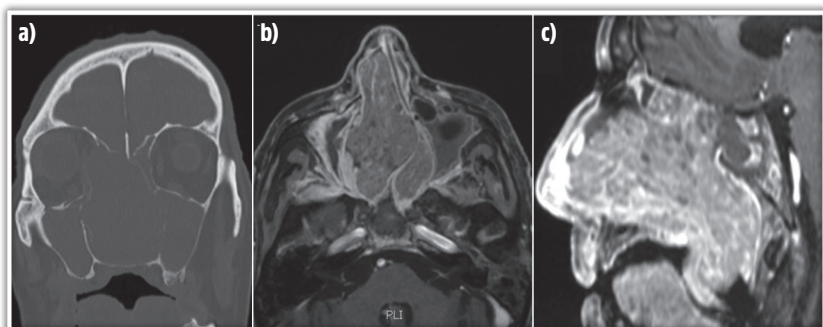
## Case description

### Clinical

- A 72 years-old male patient was referred to our tertiary center with a one-year history of progressive nasal obstruction and recurrent epistaxis. A nasal mass obstructing both vestibules was identified on physical examination.

### Imaging

- CT Scan (Figure 1a):
  - A contrast-enhancing mass obstructing both nasal cavities, predominantly on the right side was identified.
  - Lateralization of both maxillary walls and lamina papyracea was seen as well as erosion of the vomer and skull base (cribriform plate and sphenoid roof).
- MRI (Figure 1b,c):
  - The lesion measured 8.9 cm × 6.4 cm × 8.7 cm.
  - It was hypointense in T1, slightly hyperintense in T2 and showed a moderate gadolinium enhancement.
  - There was no intracranial extension or perineural involvement.
  - The mass filled the sphenoid sinus and was in close relation to the right carotid artery.
- PET CT:
  - No regional or distant metastasis were identified.



**Figure 1.** Coronal CT imaging of the invasive tumor (a). Axial (b) and sagittal (c) T1 nasopharyngeal MRI with gadolinium and fat saturation.

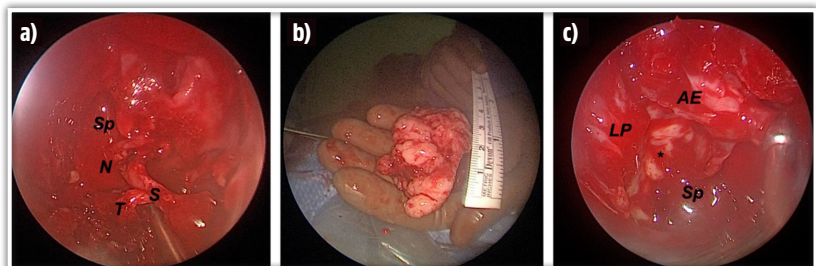
### In Office Biopsy–Pathology

- Pathologic assessment suggested an HPV-related multiphenotypic sinonasal carcinoma, with positivity for p16 and SOX-10 markers on immunohistochemistry.
- The presence of **HPV-18 DNA** was confirmed with a polymerase chain reaction analysis.

**The tumor was staged cT4N0M0 and tumor board committee recommended surgery and adjuvant radiotherapy.**

### Surgery

- Combined endoscopic and open approaches were initially planned.
- Bilateral tumor debulking was performed with a microdebrider and endoscopic forceps.
- Nasal cavity structures were thinned and lateralised by the tumor, but **all the mucosa and anatomical boundaries were preserved**.
- The mass was **pedicled on the posterior septum** and a posterior septectomy was therefore performed (Figure 2a). The tumor was removed via the oropharynx and oral cavity (Figure 2b).
- The **right optic-carotid recess was exposed** in the sphenoid sinus, but no sign of invasion was present (Figure 2c).
- Peripheral margins were sampled on the nasal septum, sphenoidal rostrum, sphenoid sinus and posterior middle and superior turbinates bilaterally. All per-operative margins were negatives.
- The posterior cribriform plate and sphenoid roof were reconstructed using a large fascia lata graft.



**Figure 2.** Posterior septal tumor attachment, Septum (S), Sphenoid sinus (Sp), Tumor (T), Nasopharynx (N) (a). Nasopharyngeal part of the tumor, removed (b). Right lamina papyracea (LP), anterior ethmoidal artery (AE) and optic-carotid recess (\*) (c).

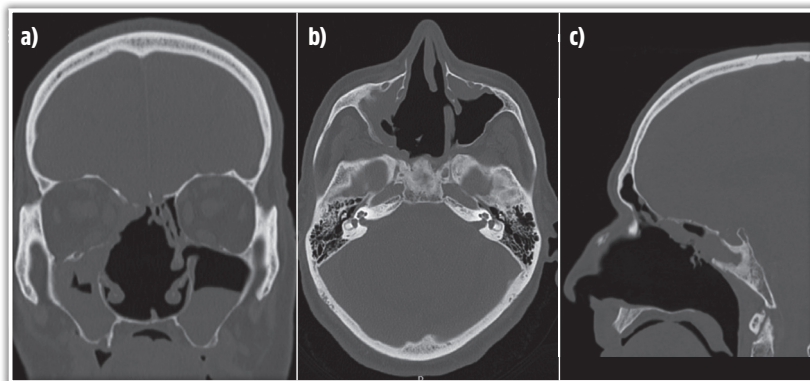
**The mass was completely removed all endoscopically.**

### Final pathology

- HMSC was confirmed on final pathologic analysis.
- Angiolymphatic invasion was identified (4 capillary invasion).
- All margins were negative and no perineural invasion was found.

### Post-operative imaging and radiotherapy

- Adjuvant postoperative radiotherapy was deferred to 10 weeks following surgical resection because of pre-irradiation dental extractions. The treatment was stopped due to ophthalmologic complications.
- A 2-months post-operative CT scan was negative for residual or recurrent tumor (Figure 3).
- Endoscopic examination was completely normal four months postoperatively.



**Figure 3.** A CT scan performed two months after surgery. Coronal (a), axial (b) and sagittal (c) views. Cranial base thickening and sphenoid opacification was secondary to the fascia lata reconstruction.

## Discussion

- HMSC is an exceedingly rare diagnosis, with fewer than a hundred cases described.
- The case described herein represents the largest tumor volume described in the English literature to this date.
- The nasal cavity is the most frequent anatomic subsite for HMSC, but this lesion can also be found in the tonsils, breasts and vaginal mucosa [6-8].
- On imaging, HMSC typically presents as a voluminous expansile sinonasal mass and remodeling of sinonasal structures as opposed to aggressive invasion seems to be a hallmark of HMSC [2, 5, 9, 10].
- Surgical management of HMSC has been the mainstay of treatment in previously published cases with or without adjuvant radiotherapy [1].
- The pedicled nature of the lesion as well as absent true invasion of structures makes for a paradoxical radiological and surgical correlation. Endoscopic surgical resections may therefore be more favorable than expected [2, 10].
- HMSC may also be a challenging diagnosis due to its overlapping histological features with other tumors such as AdCC or adenosquamous and basal cell carcinomas.
- Immunohistochemistry and proof of HPV-DNA are the cornerstones in the diagnosis of HMSC.
- While the vast majority of HMSC are associated with HPV-33, we present the second case of HMSC associated with HPV 18 [11, 12].

## Conclusion

- HMSC is a novel diagnosis in the vast spectrum of sinonasal malignancies that may be challenging for clinicians and pathologists.
- Our case report adds to literature by deepening the current knowledge of the clinical behavior as well as the paradoxical correlation between radiological and surgical findings of HMSC.
- Skull base surgeons must be familiar with this diagnosis as it may influence surgical management.

## References

Other references available upon request

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