

Content available at: <https://www.ipinnovative.com/open-access-journals>

IP Journal of Otorhinolaryngology and Allied Science

Journal homepage: <https://www.joas.co.in/>

## Case Report

# Sino nasal undifferentiated carcinoma – A diagnostic conundrum

Kamalpreet Singh<sup>1\*</sup>, Shraddha Sasi<sup>2</sup>, Poonaam Raj<sup>1</sup>, Amrindarjeet Kour<sup>3</sup>

<sup>1</sup>Dept. of ENT, Armed Forces Medical College Pune, Maharashtra, India

<sup>2</sup>Dept. of ENT, Military Hospital Bathinda, Bathinda, Punjab, India

<sup>3</sup>Dept. of Psychiatry, Armed Forces Medical College Pune, Maharashtra, India



## ARTICLE INFO

### Article history:

Received 10-12-2023

Accepted 15-02-2024

Available online 27-02-2024

### Keywords:

Head and neck cancer

Nose and Paranasal neoplasms

Sino nasal undifferentiated carcinoma

## ABSTRACT

Sinonasal undifferentiated carcinoma (SNUC) is a rare and highly aggressive tumour frequently arising from the maxillary sinus and nasal cavity. Here we report a 34-year-old female patient who presented with a pale polypoidal mass which bleeds on touch involving the left nasal cavity and nasopharynx. Imaging showed a multilobulated lesion involving the left nasal cavity and left paranasal sinuses with extension into the left masticator space, pterygopalatine fossa and retropharyngeal space supplied by a branch of the left maxillary artery. It suggested a possibility of a benign vascular mass lesion. The mass was surgically removed via the maxillary swing approach. Immunohistochemical analysis showed epithelial origin (AE1/AE3+, EMA+) and no reactivity for neuroendocrine, melanocytic, hematolymphoid, or mesenchymal markers. A diagnosis of SNUC was reached and the patient was given concurrent radiotherapy and chemotherapy. The patient is disease free one year post surgery. To conclude, a vigilant correlation of clinical, microscopic, and immunohistochemical characteristics is essential for early diagnosis and optimal management of SNUC.

This is an Open Access (OA) journal, and articles are distributed under the terms of the [Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License](#), which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

For reprints contact: [reprint@ipinnovative.com](mailto:reprint@ipinnovative.com)

## 1. Introduction

Sinonasal undifferentiated carcinoma (SNUC) is a high-grade epithelial tumor, first described by Frierson et al. in 1986. It is a rare malignancy without any evidence of squamous or glandular differentiation, with or without neuroendocrine differentiation.<sup>1</sup> The origin was presumed to be from the Schneiderian epithelium of the nose and paranasal sinuses. As per WHO, it is a highly aggressive carcinoma of uncertain histogenesis but with unique clinicopathological characteristics that typically presents as a local extensive disease.<sup>2</sup> The approximate incidence rate of SNUC is 0.02 per 100,000 according to data from the Surveillance, Epidemiology, and End Results (SEER) database,<sup>3</sup> Histologically, SNUC is a small round

blue cell tumour due to prominent nuclei with a high mitotic rate. It is immunohistochemically distinct from other sinonasal malignancies, such as nasopharyngeal carcinoma, neuroendocrine carcinoma, lymphoma, mucosal melanoma, and olfactory neuroblastoma.<sup>1</sup>

SNUC generally shows male predominance (2–3:1). Age of presentation ranges from the 3rd to 9th decade with the median age at presentation of 50 to 60 years.<sup>4,5</sup> It typically involves the nasal cavity, maxillary antrum, and ethmoid sinus either alone or in combination and invades other contiguous sites such as the skull base or orbit.<sup>2</sup> It usually presents at an advanced stage.<sup>6</sup> The presenting symptoms may be multiple, ranging from nasal symptoms such as nasal obstruction, epistaxis, facial pain, and headache to severe symptoms like proptosis, diplopia, impaired visual acuity and cranial nerve palsies. The duration of symptoms is relatively short, varying from weeks to months. Around

\* Corresponding author.

E-mail address: [kpsingh\\_81@yahoo.com](mailto:kpsingh_81@yahoo.com) (K. Singh).

10–30% of SNUC patients initially present with cervical lymph node metastases, while distant metastases are rare at initial presentation.<sup>7</sup>

Both computed tomography (CT) and magnetic resonance imaging (MRI) are essential for the assessment of the degree of local invasion of the tumour and the presence of cervical lymphadenopathy. Additional imaging like positron emission tomography (PET) and CT chest may be utilized to assess the presence of distant metastasis.<sup>8</sup> American Joint Committee on Cancer (AJCC) staging system is utilized for grading the severity of the disease. The management of SNUC is formidable as these endoscopic tumours lie in areas difficult to reach and cause frequent invasion of critical nearby structures. Most data in the literature establish the benefit of multimodality treatment in improving outcomes. The advent of sinus and skull base surgery over the recent years has resulted in a dramatic decrease in morbidity for selected cases and might consequently offer an improved quality of life in patients with poor prognosis.

## 2. Case Report

A 34-year-old woman presented to our centre with symptoms of left nasal obstruction, facial pain, and rhinorrhoea with recurrent epistaxis and restricted mouth opening. A physical examination showed swelling and tenderness in the left cheek, and a nasal endoscopy revealed a pale polypoidal mass occupying the left nasal cavity and extending into the right choana. The mass bled on touch. However, she displayed no ocular symptoms. A computed tomography (CT) revealed a multilobulated lesion involving the nasal cavity displacing the nasal septum to the right, nasopharynx extending into masticator space involving medial and lateral pterygoid muscles, retropharyngeal space and causing scalloping and sclerosis of the left half of base of the skull. The lesion was extending into the left infratemporal fossa eroding the left maxillary bone. The features were suggestive of a vascular mass supplied by branches of the maxillary artery of the left external carotid artery. Subcentimetric nodes were present in levels Ib, II and V bilaterally. Magnetic resonance imaging (MRI) revealed a heterogeneously hyperintense lesion in the T2 weighted and an iso-hypointense lesion in the T1 weighted image. Biopsy was not taken given the lesion being a highly vascular mass.

Immunohistochemically, the tumour cells of undifferentiated carcinoma are positive for cytokeratins (AE1/AE3) and epithelial membrane antigen and showed a high proliferation index in Ki67 immunostaining. The tumour cells were negative for Melan A, S100, LCA, and Synaptophysin with Chromogranin being focally positive. Positron emission tomography (PET) revealed remnant FDG avid soft tissue lesion in the left ethmoid sinus and nasopharynx reaching up to the skull base (maximum standardized uptake value [SUV] = 7.06) with FDG

avid cervical, axillary, mediastinal and upper abdominal nodes. The patient was treated with chemotherapy and radiotherapy. A 3-month follow-up was uneventful.

## 3. Discussion

SNUC is an exceptionally rare and aggressive tumour arising from the Schneiderian epithelium lining the sinonasal tract. Due to the absence of any pathognomonic feature associated with SNUCs, the diagnosis appears challenging. Our case had a clinical picture similar to a benign neoplasm likely nasopharyngeal angiofibroma. However, age and gender were contradicting factors. Most often, these tumours are diagnosed at an advanced stage. Orbit, skull base and intracranial extension are commonly observed. Musy et al. demonstrated that 50% of patients have a dural invasion at the time of presentation and 30% have an orbital invasion<sup>9</sup>. In another study, all 19 patients with SNUC had stage T4, 14 patients (74%) had dural involvement, 4 (21%) had cavernous sinus invasion, and 12 patients (63%) had orbital apex involvement. But our case had no intracranial or intraorbital extension.<sup>10</sup>

Immunohistochemically, the tumour cells are immunoreactive for pan-cytokeratins and simple keratins (CK7, CK8 and CK19) and negative for CK4, CK5/CK6 and CK14. Less than half of the cases have been reported to be positive for epithelial membrane antigen, neuron-specific enolase, or p53.<sup>5</sup>

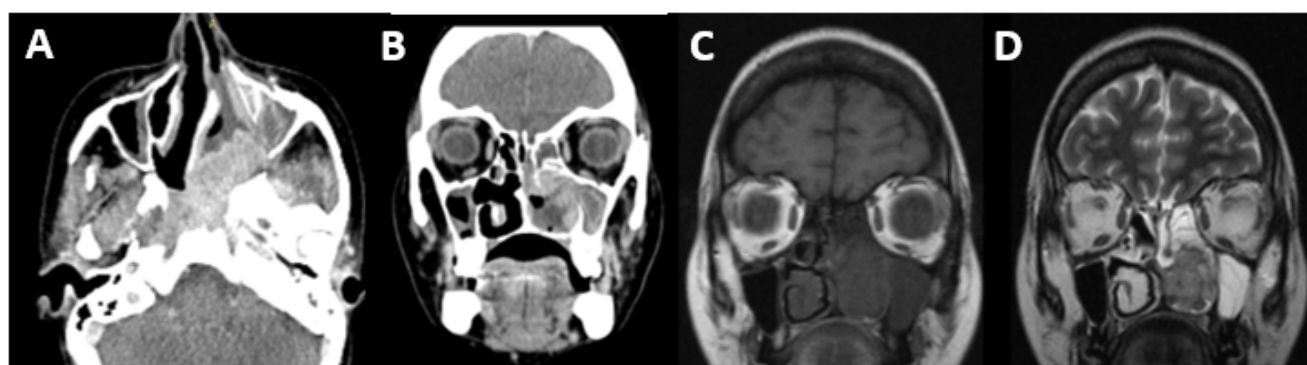
Craniofacial resection followed by radiation or chemoradiation therapy is considered the standard treatment for SNUC, for advanced-stage and close-tumour resection margins. Many studies also reported that the use of a three-modality treatment would provide a better local control compared to the two-modality approach.<sup>11,12</sup> SNUC carries a dismal prognosis. According to a recent large SEER database analysis, 5-year and 10-year survival was found to be 34.9% and 31.3% respectively with a median survival of 22.1 months.<sup>3</sup>

## 4. Conclusion

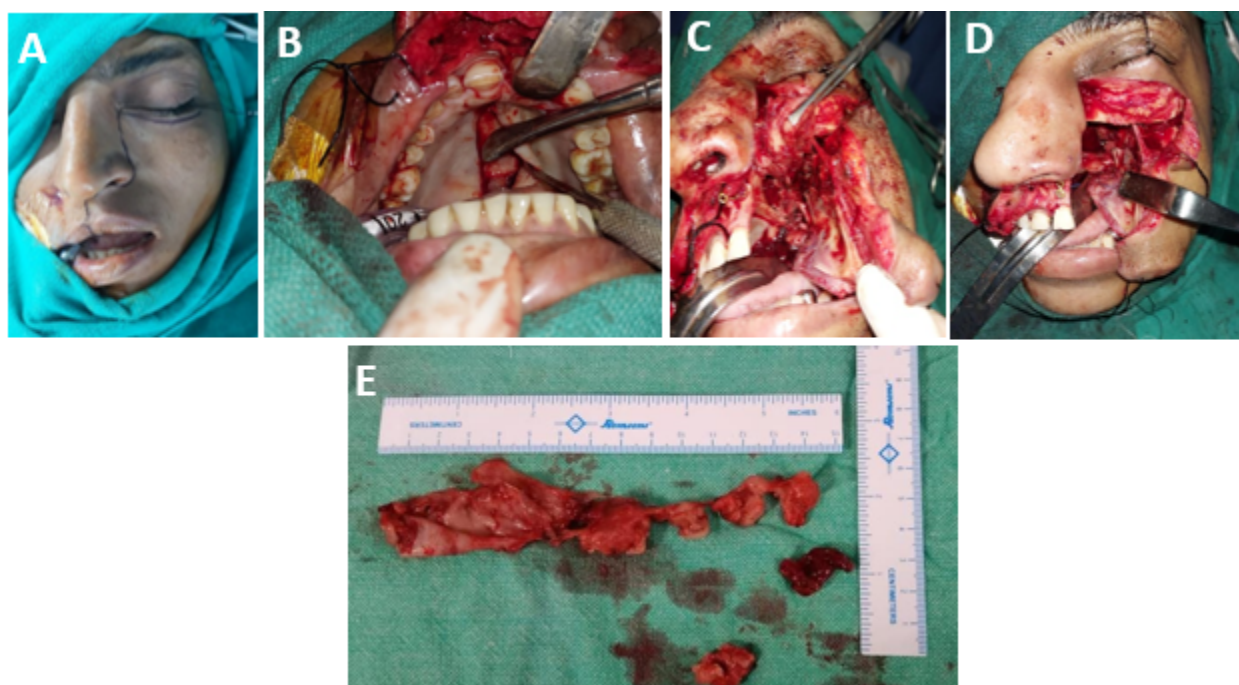
Sinonasal Undifferentiated Carcinoma is mostly a diagnosis of exclusion. Due to the likelihood of a nonspecific finding at the time of presentation and the rarity of this tumour, the diagnosis runs the risk of being delayed. Special attention should be made to differentiate SNUC from other sinonasal malignancies to guide the appropriate treatment. SNUC has a poor prognosis with high rates of locoregional recurrence and a tendency to metastasize. The recent advances in radiation techniques, chemotherapeutic agents, and promising targeted therapy have a potential role in improving the outcome.

## 5. Source of Funding

None.



**Figure 1:** Axial computed tomogram showing a brightly enhancing mass lesion extending into the left infratemporal fossa(A). Coronal computed tomogram showing the lesion involving the left nasal cavity left maxillary and ethmoid sinuses (B). MRI shows a heterogenous signal intensity in the T1 (C) and T2 (D) weighted images. a maxillary swing approach anaesthesia to remove the maxillary, ethmoid and sphenoid sinuses; left infratemporal fossa and attached to



**Figure 2:** Surgical removal of the mass through a Modified Weber Fergusson incision with Dieffenbach extension and mid-palatal incision (A, B). Mass removed in toto (C-E).

## 6. Conflict of Interest

None.

## References

1. Frierson HF, Mills SE, Fechner RE, Taxy JB, Levine PA. Sinonasal undifferentiated carcinoma. An aggressive neoplasm derived from Schneiderian epithelium and distinct from olfactory neuroblastoma. *Am J Surg Pathol.* 1986;10(11):771–80.
2. Frierson HF. Sinonasal undifferentiated carcinoma,” in World Health Organization classification of tumours. *Pathol Genetics.* 2005;28(4):1–19.
3. Chambers KJ. Incidence and survival patterns of sinonasal undifferentiated carcinoma in the United States. *J Neurol Surg B Skull Base.* 2015;76(2):94–100.
4. Jeng YM. Sinonasal undifferentiated carcinoma and nasopharyngeal-type undifferentiated carcinoma: two clinically, biologically, and histopathologically distinct entities. *Am J Surg Pathol.* 2002;26(3):371–7.
5. Cerilli LA, Holst VA, Brandwein MS, Stoler MH, Mills SE. Sinonasal undifferentiated carcinoma: immunohistochemical profile and lack of EBV association. *Am J Surg Pathol.* 2001;25(2):156–63.
6. Zielinski V. Management of sinonasal undifferentiated carcinoma with intracerebral invasion: Clinical experience at a single institution and review of the literature. *Ear Nose Throat J.* 2016;95(1):23–31.
7. Tanzler ED, Morris CG, Orlando CA, Werning JW, Mendenhall WM. Management of sinonasal undifferentiated carcinoma. *Head Neck.* 2008;30(5):595–604.

8. Mendenhall WM, Mendenhall CM, Riggs CE, Villaret DB, Mendenhall NP. Sinonasal undifferentiated carcinoma. *Am J Clin Oncol*. 2006;29(1):27–31.
9. Musy PY, Reibel JF, Levine PA. Sinonasal undifferentiated carcinoma: the search for a better outcome. *Laryngoscope*. 2002;112(8):1450–5.
10. Gray ST. Treatment outcomes and prognostic factors, including human papillomavirus, for sinonasal undifferentiated carcinoma: a retrospective review. *Head Neck*. 2015;37(3):366–74.
11. Mourad WF. Trimodality management of sinonasal undifferentiated carcinoma and review of the literature. *Am J Clin Oncol*. 2013;36(6):584–92.
12. Al-Mamgani A, Rooij PV, Mehilal R, Tans L, Levendag PC. Combined-modality treatment improved outcome in sinonasal undifferentiated carcinoma: single-institutional experience of 21 patients and review of the literature. *Eur Arch Otorhinolaryngol*. 2013;270(1):293–302.

## Author biography

**Kamalpreet Singh**, Associate Professor

**Shraddha Sasi**, Senior Resident

**Poonaam Raj**, Professor and HOD

**Amrindarjeet Kour**, Senior Resident

**Cite this article:** Singh K, Sasi S, Raj P, Kour A. Sino nasal undifferentiated carcinoma – A diagnostic conundrum. *IP J Otorhinolaryngol Allied Sci* 2023;6(4):119-122.