Section: Pathology

Case Series

SINONASAL CARCINOMA MASQUERADING AS FUNGAL SINUSITIS: A CASE SERIES OF RARE PRESENTATION

Savita Agarwal[1], Asttha Kapoor[2], Pinki Pandey[3], Udbhav Tewari[4]

[1]Professor, Department of Pathology, U.P.UM.S, Saifai, Etawah, Uttar Pradesh, India.
[2]Senior Resident, Department of Pathology, U.P.UM.S, Saifai, Etawah, Uttar Pradesh, India.
[3]Professor and Head, Department of Pathology, U.P.UM.S, Saifai, Etawah, Uttar Pradesh, India.
[4]Senior Resident, Department of Psychiatry, U.P.U.M.S, Saifai, Etawah, Uttar Pradesh, India

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Corresponding Author:
Dr. Asttha Kapoor,
Email: docasttha@rediffmail.com

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ABSTRACT

Sinonasal undifferentiated carcinoma (SNUC) is a rare type of cancer that affects the nasal cavity and paranasal sinuses. It has a very low incidence of 0.02 per1 lakh. SNUC is a highly aggressive, undifferentiated anaplastic carcinoma, without obvious squamous, glandular, neuroectodermal, mesenchymal, melanocytic or other lines of differentiation. As there are few cases with <400 cases documented in the literature since the original series that have been reported, the best course of treatment has not been thoroughly evaluated. Because of the high rate of distant metastases and loco-regional recurrence, SNUC is treated with intensive multimodality therapy that includes adjuvant treatment (i.e., chemotherapy and radiation).

INTRODUCTION

Sinonasal undifferentiated carcinoma (SNUC) is an uncommon and extremely aggressive cancer for which there are no specific treatment guidelines or reliable stage-based survival statistics. With 5-year survival rates in the literature ranging from 20 to 63%, overall SNUC mortality rates are significant.[1] Early in the course of the disease, neighbouring structures are regularly invaded and eroded by SNUC, which causes great local destruction.[2] Furthermore, it typically manifests at stage IV of the American Joint Committee on Cancer (AJCC).[1,3]

Although there are many contributing factors to the severe state of the disease at presentation, one important aspect is the delay in diagnosis, as early SNUC symptoms are more often mistaken for benign causes. A patient with a locally advanced SNUC frequently shows up for assessment with sinusitis, face pressure, or congestion of the nose. The enormous potential space of the sinuses provides for unrestricted initial growth, which helps to explain the tumor's high early T stage.[4]

Frierson (1986) was the first to report on sinonasal undifferentiated carcinoma (SNUC), an unusual malignancy of the paranasal sinuses and nasal cavity.[3] Histological investigation may reveal striking similarities between SNUC and olfactory neuroblastoma. It is necessary to distinguish olfactory neuroblastoma from SNUC because of its significantly superior prognosis. It can be difficult to confirm the light microscopic diagnosis of SNUC because there are several possible microscopic differential diagnoses that include neuroendocrine carcinoma, malignant melanoma, olfactory neuroblastoma, rhabdomyosarcoma, and undifferentiated nasopharyngeal carcinoma (lymphoepithelioma). It is possible to distinguish sinonasal undifferentiated carcinoma from several other neoplasms by comparing their clinical, immunohistochemical, light microscopy, and ultrastructural features.[5] Although vigorous multimodal therapy may be the best line of action for treating this neoplastic disease locally. The purpose of this case series is to present cases of SNUC and to review the current concepts of management for this entity.

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CASE SERIES

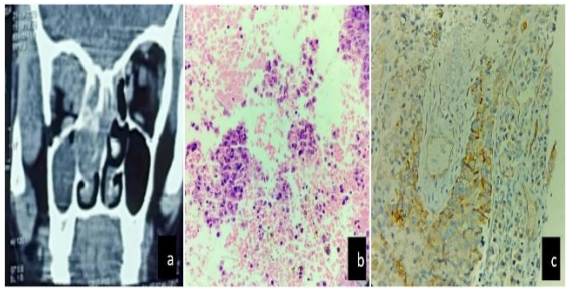

Figure 1: a. NCCT scan showing heterogeneously enhancing mass lesion with its epicenter in bilateral ethmoid sinuses and extending posteriorly to involve bilateral sphenoid sinuses. There is destruction of medial bony wall of right orbit with proptosis and lateral displacement of the globe with impingement of the mass on right optic nerve at the apex. B. Section showing Poorly differentiated carcinoma H& E (X400). C. IHC image of PAN CK (X400)

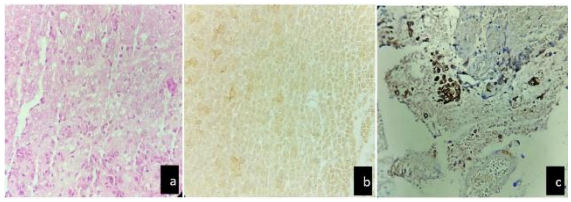

Figure 2: a Section showing poorly differentiated carcinoma H & E (X400). B. IHC image of PAN CK (X400). C. IHC image of CK 7 (X100)

3 patients who presented to the out-patient department of our institute with common complaints of sinusitis and congestion of nose on detailed workup and evaluation were finally diagnosed as Sinonasal undifferentiated carcinoma on the basis of histopathology and immunohistochemistry. Case details, examination findings, radiological findings, histopathological and immunohistochemical findings of the patients are tabulated in [Table 1].

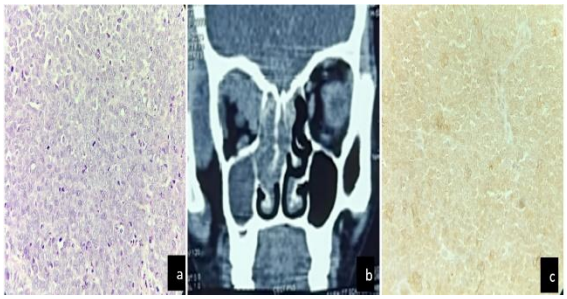

Figure 3: a Section showing poorly differentiated carcinoma with brisk mitosis H & E (X400) (X400). b NCCT scan showing soft tissue density areas involving right nasal cavity, right ethmoid, sphenoid sinus and right orbit. c IHC image of PAN CK (X400).

Table 1: Details of cases with complete workup.

Features	Case-1	Case-2	Case-3
Age/Sex	24year/Female	60year/Male	20year/Female
Chief complaints	Nasal bleeding, Nasal Obstruction x 3 months, Protrusion of right eye x 2 months Progressive reduction in visual acuity x 3 weeks	Nasal bleeding, Left eye proptosis headache x 1 month.	Nasal bleeding, Nasal Obstruction x 3 months.
Examination findings	Nasal Cavity: Reddish and friable mass stuffing right nasal cavity which was firm in consistency, bled on probing and led to left side deviation of nasal septum. Eye: Lateral and outward protrusion of right eye with restricted eye movement and reduced visual acuity.	Nasal Cavity: Reddish and friable mass stuffing left nasal cavity which was firm in consistency, bled on probing and led to right side deviation of nasal septum. Eye: Lateral and outward protrusion of left eye with restricted eye movement.	Nasal Cavity: Reddish white and friable mass stuffing right nasal cavity which was firm in consistency, did not bleed on probing and led to right side deviation of nasal septum. Eye: Lateral and outward protrusion of right eye with restricted eye movement.
CT Findings of Paranasal sinuses and orbit	Heterogeneously enhancing soft-tissue mass lesion with bony erosion and extension into right orbit, bilateral nasal cavities, right frontal sinus, bilateral sphenoid sinuses and right optic nerve. Fig.1a	Soft tissue density areas involving left nasal cavity, left ethmoid, sphenoid sinus and left orbit with invasion of base of skull at frontal lobe region. Significant bony erosion was seen involving adjacent bones	Soft tissue density areas involving right nasal cavity, right ethmoid, sphenoid sinus and right orbit. Fig. 3a
Clinical differential diagnosis	Fungal Sinusitis Nasal mass	Fungal sinusitis Nasal mass	Fungal Sinusitis Nasal mass
Histopathology	Poorly differentiated carcinoma with large areas of necrosis Fig. 1b	Poorly differentiated carcinoma with partly ulcerated mucosa. Fig. 2a	Poorly differentiated carcinoma with brisk mitosis. Fig. 3b
Immunohistochemistry	Pan CK +ve Fig. 1c CK 7 +ve Vimentin -ve Leukocyte common antigen (LCA) -ve S-100 -ve	Pan CK +ve Fig. 2b CK 7 +ve Fig. 2c Vimentin -ve Leukocyte common antigen (LCA) -ve S-100 -ve	Pan CK +ve Fig. 3c Vimentin +ve CD56 +ve NSE +ve Leukocyte common antigen (LCA) -ve S-100 -ve

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Final Diagnosis	Sinonasal undifferentiated carcinoma	Sinonasal undifferentiated carcinoma	Sinonasal undifferentiated carcinoma

DISCUSSION

First identified by Frierson et al. in 1986, sinonasal undifferentiated carcinoma is an aggressive tumor that differed clinicopathologically from other poorly differentiated cancers of the sinuses and nasal cavity.[3] It can afflict people of any age, from the third to the ninth decades, with male predominance (2–3:1), however it most frequently manifests in the sixth decade.[6] Nasopharyngeal carcinoma of undifferentiated type typically presents as a polypoidal mass extending into lateral wall of nasopharynx, superior posterior wall. Clinically patient presents as cervical neck mass, nasal obstruction, nasal discharge, epistaxis, otalgia, serous otitis media, hearing loss and headache. The tumor cells have indistinct borders with a round nucleus, fine chromatin, prominent nucleoli and scant cytoplasm. Immunohistochemistry demonstrates staining for PAN CK +, CK 7-, LCA-.[7]

It can be challenging to distinguish SNUC from neuroendocrine carcinoma and olfactory neuroblastoma because, while some SNUC cells express neuroendocrine markers like chromogranin and NSE, S-100 is absent, and there is no sign of glandular differentiation or rosettes. Necrosis and cytological atypia are also more prevalent in SNUC. In SNUC, neuroendocrine indicators are typically localized and lack ultrastructural evidence of neurosecretory granules.[8] Squamous cell carcinomas that are poorly differentiated typically feature distinct cytokeratin patterns and little patches of visible squamous differentiation or keratinization. Immunohistochemically, many SNUCs stain for both epithelial markers and all are positive for either EMA or cytokeratin. Roughly 50% of the tumors exhibit positive results for NSE, but the majority do not exhibit S-100 immunoreactivity and are vimentin-negative.[5] Multiple paranasal sinuses may be affected by these tumors, with significant sinus wall damage. The majority of these characteristics align with the outcomes in our instance. A heterogeneous mass lesion that destroys bone and invades neighboring tissues, such as the orbits, paranasal sinuses, and anterior cranial fossa, is the radiologic characteristic of SNUC.[2] In our cases, there was clear bone loss and involvement of nearby structures. The optimal course of treatment has not been thoroughly assessed because there are only a few cases that have been documented. Nonetheless, the tumor is typically surgically removed as part of the treatment.

Extensive craniofacial excision usually entails maxillectomy, orbital exenteration, and on rare occasions, neurosurgery intervention. Both distant metastasis and local regional recurrence are common in SNUC patients. Aggressive multimodality therapy, such as surgical resection and adjuvant therapy (i.e., radiotherapy and chemotherapy), are used in the treatment of SNUC. Etoposide, cisplatin, and carboplatin are among the chemotherapy drugs utilized.[9] With SNUC, the prognosis is not good, and death frequently happens soon after the diagnosis. Patients with SNUC treated with radiotherapy alone had a median survival of only 4 months, according to the original report by Frierson et al,[3] demonstrated that an intensive multimodality approach improved the survival (median 53.6 months) results for SNUC. According to a meta-analysis by Chambers et al,[10] the National Cancer Institute's surveillance, epidemiology, and end outcomes (SEER) program yielded a total of 318 cases of SNUC in the United States between 1973 and 2010. The five-year relative survival rates for patients who underwent radiation, surgery, or both were 36.0, 39.1%, and 38.7%, respectively. 41.9 months was the median survival for radiotherapy and surgery combined. The median survival for the years 1973–1986 and 1986–2010 was 14.5 and 23.5 months, respectively; this finding confirms the effectiveness of radiation therapy and surgery for survival and points to a recent trend of increased survival. The cure rate for SNUC is still incredibly low, even with advancements in chemoradiation therapy and anterior skull base surgery.[11,12]

CONCLUSION

Since early care may enhance survival and outcome, it is imperative to detect and differentiate this distinct entity from other paranasal sinus tumors due to its aggressive activity. Histopathological diagnosis is must to differentiate and categorise these tumors.

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