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Sinonasal Undifferentiated Carcinoma—A Case Report

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Abstract

Sinonasal tumors with neuroendocrine differentiation are uncommon. They arise from schneiderian epithelium. The authors reported a case in a fifty-six years old female patient who came with complaints of headache and epistaxis. Differential diagnosis and review of literature are presented in detail. This case is presented in view of its rarity.

Keywords

Sinonasal Undifferentiated Carcinoma, Sinonasal Neuro Endocrine Carcinoma, Schneiderian Epithelium

1. Introduction

Sinonasal carcinomas comprise less than 1% of all neoplasms and 3% of those of the upper aero-digestive tract. Primary sinonasal neuro endocrine tumors show a varied histomorphological spectrum ranging from esthesioneuroblastoma (ENB), sinonasal undifferentiated carcinoma (SNUC), sinonasal neuroendocrine carcinoma, and small cell undifferentiated carcinoma. Sinonasal undifferentiated carcinoma is a highly aggressive neoplasm, and it has to be differentiated from less aggressive sinonasal tumors. They differ in their cell of origin, degree of neuroendocrine differentiation and biologic behavior [1].

Case history: Patient came with complaints of headache since one year, blurring of vision, vertigo, mass over nasal bridge which is hard and tender since two months, and epistaxis since one month. She was a known diabetic and hypertensive. She suffered with right side hemiparesis, recovered after one month. CT scan of paranasal sinuses showed a mass lesion in frontal sinus with extension into bilateral ehmoidal sinuses, left orbit and nasal cavity (**Figure 1**). Flexible nasopharyngoscopy showed a mass in right nasal cavity looking like an inverted papilloma. Left nasal cavity showed a small polypoidal mass. Left ethmoidal sinus was opened and mass removed.

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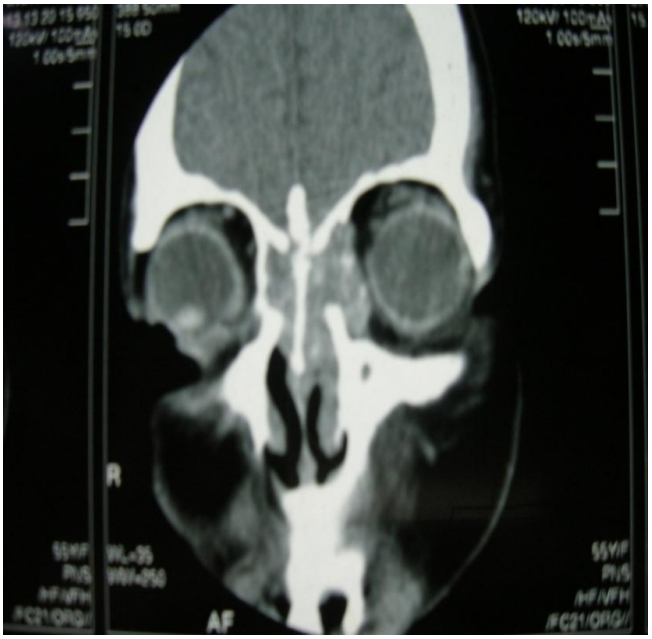

Figure 1. CT scan picture of paranasal sinuses showing extension into paranasal sinuses.

Gross features: We received multiple grey brown bits measuring 2.5 cm × 1.5 cm × 1 cm. And the entire tissue was submitted for histopathology.

Microscopy: Tumor cells are arranged in diffuse sheets with overlying pseudostratified ciliated columnar epithelium. Tumor cells are small in size arranged in sheets with marked pleomorphism; nucleus showed coarse chromatin and prominent nucleoli (**Figure 2** and **Figure 3**). Areas of necrosis and vascular proliferation are seen. Mitotic figures are plenty. We have considered differential diagnosis of sinonasal neuroendocrine carcinoma and sinonasal undifferentiated carcinoma. We have ruled out esthesioneuroblastoma because there was no neuropil. We have done IHC markers for EMA (epithelial membrane antigen), synaptophysin. Tissue was positive for EMA and negative for synaptophysin (**Figure 4** and **Figure 5**).

2. Discussion

Sinonasal tumors with neuroendocrine differentiation are uncommon tumors with considerable overlap of histological features. Cell of origin may be related to both schneiderian and olfactory epithelia.

Four histologic phenotypes are described [1].

- Esthesioneuroblastoma (ENB).
- Sinonasal neuroendocrine carcinoma (SNEC).
- Sinonasal undifferentiated carcinoma (SNUC).
- Small cell carcinoma (SmCC).

In this spectrum esthesioneuroblastoma is at the most differentiated end and small cell carcinoma at the least differentiated end.

Sinonasal neuroendocrine carcinoma resides within the less differentiated end of the spectrum of neuroendocrine tumors relative to olfactory neuroblastoma. Sinonasal neuroendocrine carcinoma is a cellular tumor lacking the fibrillary background of olfactory neuroblastoma. Cytologically cells are larger with more cytoplasm, coarse chromatin and larger nucleoli. There can be overlapping of features [2].

The term sinonasal undifferentiated carcinoma is coined by Frierson to describe a group of undifferentiated, aggressive tumors with no obvious differentiation. Most tumors arise in the nasal cavity and can extend into ethmoid and antra [3]. Presentation with orbital or intracranial tumor presentation is common. Abdul Wadood *et al*. have reported a case with contralateral sinus involvement [4]. Exposure to heavy metals, coal mining, working in chemical industry are supposed to the etiological factors. Epstein Barrvirus has been identified in

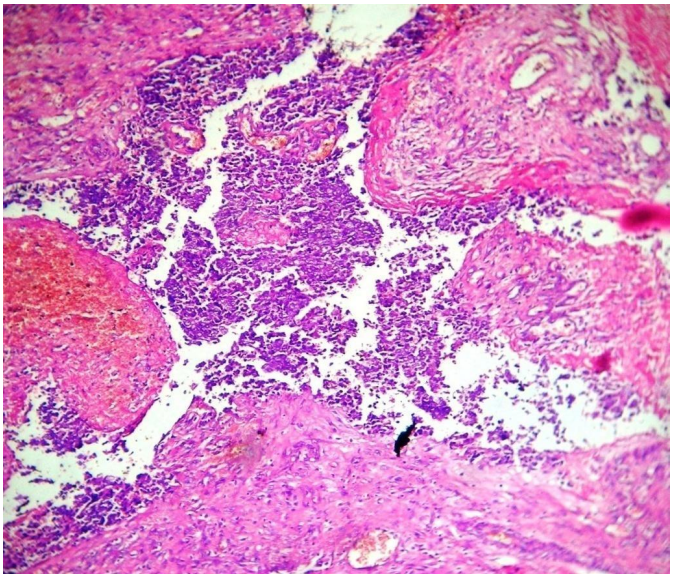

Figure 2. 10 × 10 view, H & E stain showing nests of tumor cells with areas of necrosis.

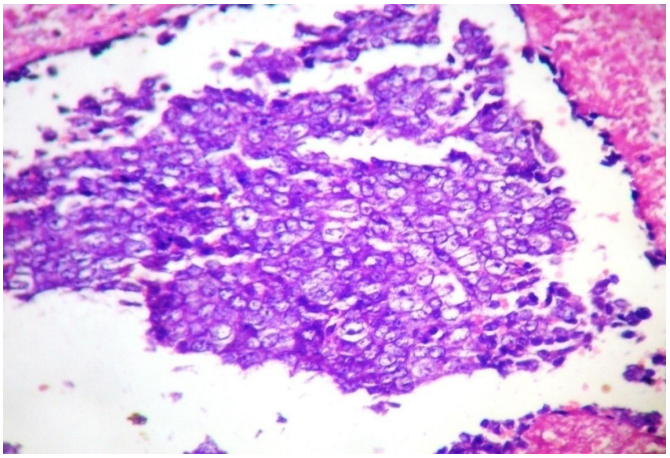

Figure 3. 10 × 40 view, H & E stain, tumor cells with prominent nucleolus.

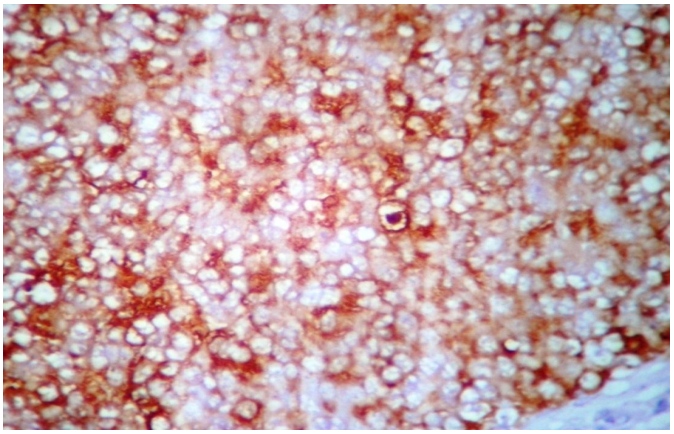

Figure 4. 10 × 40 view, IHC positive for EMA.

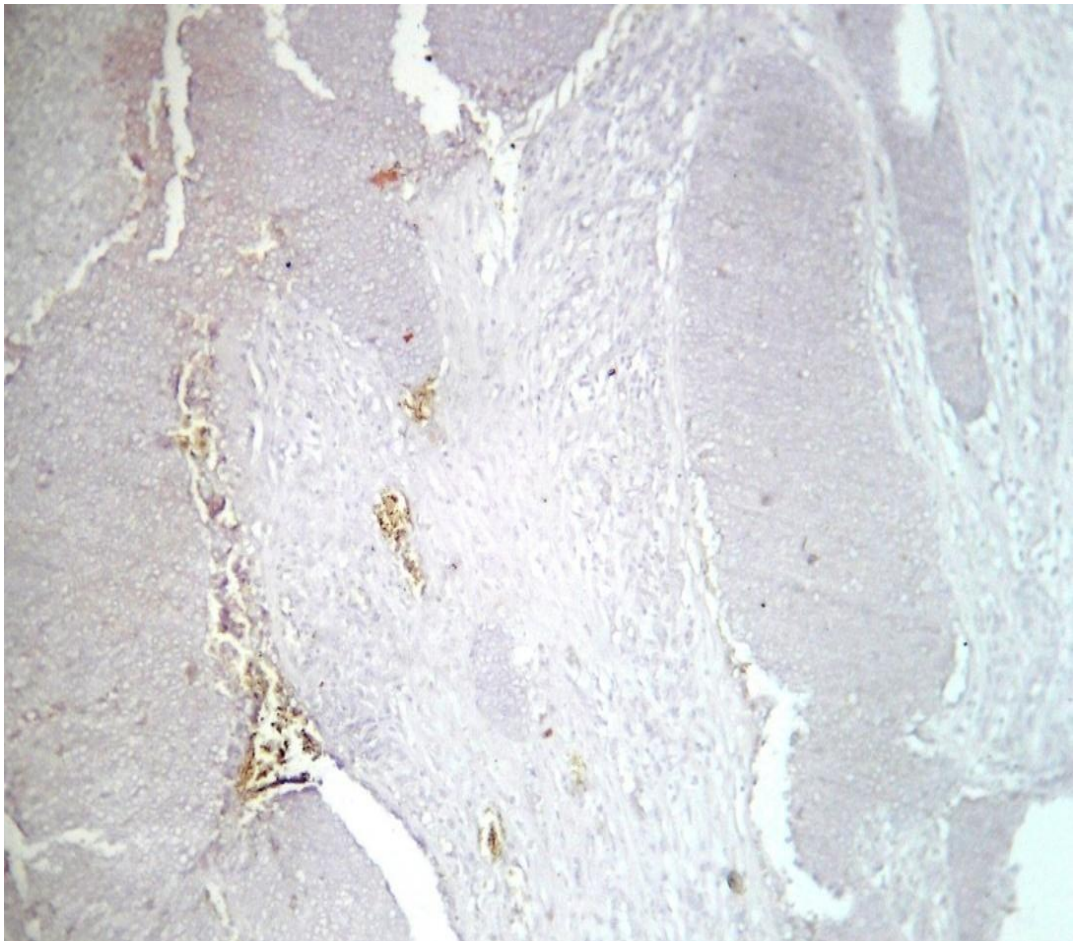

Figure 5. 10 × 40 view, IHC negative for synaptophysin.

Asian and Italian patients but not in other western patients [3] [5]. Deletion of the retinoblastoma gene has been implicated. Sinonasal undifferentiated carcinoma is a diagnosis of exclusion. High nuclear cytoplasmic ratio, either small cells or large cells, coarse chromatin, prominent nucleoli, numerous mitoses, necrosis are the features. Surface mucosal *in situ* carcinoma can be seen. Osteoclast like giant cells, spindle cells can be rarely seen. The limited electron microscopic evidence of neuroendocrine differentiation aids in distinguishing sinonasal neuroendocrine carcinoma from sinonasal undifferentiated carcinoma.

Immunohistochemistry; sinonasal neuroendocrine carcinoma is synaptophysin +ve, epithelial membrane antigen +ve where as sinonasal undifferentiated carcinoma is synaptophysin −ve, epithelial membrane antigen +ve. Natural history and biological behaviour varies in this group of tumors. The recommended treatment consists of an aggressive combination of craniofacial resection, adjuvant radiotherapy and chemotherapy. Even then dismal outcomes are common [2] [6].

In the present case the tumor showed cells with high nuclear cytoplasmic ratio, coarse chromatin, prominent nucleoli, and areas of necrosis. Immunohistochemistry showed negativity for synaptophysin and positivity for EMA. Because there was pleomorphism, areas of necrosis, no neuropil and it was extending into ethmoid sinus it was given “Hyams” grade 4 and “Kadish” stage C (**Table 1**, **Table 2**). Hence the histomorpho diagnosis coupled with grading and staging of the tumors is important in the prognostication of these tumors. In the present case functional endoscopic sinus surgery (FESS) was done and post-operatively external radiation of 6250 rads was given followed by 5 cycles of chemotherapy. The drugs used were dacarbazine, adriamycin. The patient succumbed to disease after seven months. Miyomoto *et al*. studied a series of cases in which one case with Kadish stage C and one case with Hyams grade 4 survived for more than five years [7]. But in the present study even with aggressive treatment patient succumbed after 7 months.

Table 1. Hyams histological grading.

gr4	gr3	gr2	gr1	Histologic criteria
−	−	+	++	Lobular architecture
−	+/−	+	++	Neuropil
−	+/−	+/−	+/−	Rosettes
++	+	−	−	Necrosis
+++	++	+	−	Pleomorphism

Table 2. Morita modification of Kadish staging.

Tumor involving nasal cavity	Stage A
Involving paranasal sinuses	Stage B
Extending beyond sinuses to involve orbit	Stage C
Metastasis to cervical nodes or distant metastasis	Stage D

3. Conclusion

Finally it was signed out as sinonasal undifferentiated carcinoma, Hyams grade: grade 4, Kadish stage C (**Table 1**, **Table 2**).

References

[1] Menon, S., Pai, P., Sengar, M., Aggarwal, J.P. and Kane, S.V. (2010) Sinonasal Malignancies with Neuroendocrine Differentiation: Case Series and Review of Literature. *Indian Journal of Pathology & Microbiology*, **53**, 28-34. http://dx.doi.org/10.4103/0377-4929.59179

[2] Brandwein-Gensler, M. (2006) Sinonasal and Nasopharyngeal Surgical Pathology in Silverberg's Principles and Practice of Surgery and Cytopathology. 4th Edition, Elsevier, Amsterdam, 787-822.

[3] Wenig, B.M. and Pilch, B.Z. Tumors of the Upper Respiratory Tract in Diagnostic Histopathology of Tumors. In: Fletcher, C.D.M., Ed., 2nd Edition, Boston, 111-116.

[4] Mohammed, A.W., Bakshi, J. and Nada, R. (2013) Sinonasal Undifferentiated Carcinoma with Contralateral Cavernous Sinus Involvement—A Rare Presentation. *Current Research in Microbiology and Biotechnology*, **1**, 58-61.

[5] Jones, A.V., Robinson, I. and Speight, P.M. (2005) Sinonasal Undifferentiated Carcinoma: Report of a Case and Review of Literature. *Oral Oncology Extra*, **41**, 299-302. http://dx.doi.org/10.1016/j.ooe.2005.07.007

[6] Xu, C.C., *et al*. (2013) Sinonasal Undifferentiated Carcinoma: The Alberta Experience and Literature Review. *Journal of Otolaryngology—Head and Neck Surgery*, **42**, 2. http://dx.doi.org/10.1186/1916-0216-42-2

[7] Miyomoto, R.C., *et al*. (2000) Esthesioneuroblastoma and Sinonasal Undifferentiated Carcinoma: Impact of Histological Grading and Clinical Staging on Survival and Prognosis. *The Laryngoscope*, **110**, 1262-1265. http://dx.doi.org/10.1097/00005537-200008000-00007

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