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CASE REPORT

Primary Small Cell Carcinoma of Nasal Cavity Presenting as Unilateral Blindness

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ABSTRACT

Primary sinonasal small cell carcinomas are extremely rare lesions. We present a case of 29-year-old male patient, who initially presented with recurrent right sided nasal obstruction and occasional epistaxis, but came to medical attention when he developed progressive deterioration of vision in ipsilateral eye. Radiological examination showed a tumor with epicenter in right nasal cavity with extensive local infiltration. The histopathology picture and immunohistochemistry profile of the tumor showed it to be a primary small cell neuroendocrine tumor. Extensive imaging studies revealed no other site of primary. He was treated with induction chemotherapy followed by concomitant chemoradiotherapy. Complete clinical response was seen and the patient has been disease free since last 20 months. The report discusses the diagnostic criteria which differentiates this extremely rare but aggressive tumor from a much more common and more indolent tumor, esthesioneuroblastoma.

Keywords: Nasal cavity, Paranasal sinuses, Neuroendocrine carcinoma, Small cell carcinoma.

INTRODUCTION

Extrapulmonary small cell carcinomas (EPSCCs) are uncommon malignant neoplasms with a reported incidence of 0.1 to 0.4%.[1] While primary sites documented may include esophagus, salivary glands, gastrointestinal tract (including small intestine and large intestine), pancreas, larynx, cervix uteri, uterus, urinary bladder, prostate, breast and lacrimal gland, primary sinonasal small cell carcinoma (SmCC) are extremely rare. This tumor was first reported in 1965, and since then there have been only 61 documented cases in the literature.[2] We present a case of 29-year-old male patient who presented with progressively increasing right-sided nasoethmoid mass lesion and progressive blindness in ipsilateral eye . The histopathology of this rare tumor and difficulty in treatment strategy are discussed.

CASE REPORT

A 29-year-old, nontobacco user, presented with one year history of recurrent right-sided nasal obstruction and a few episodes of epistaxis. Over previous 3 months period, he had developed progressive vision loss in the right eye and decreased sensation on the right side of face. On examination, he had broadening of nasion (Fig. 1). A red, friable, gelatinous, polypoid mass with a tendency to bleed was seen in the right nasal cavity. Right side pupils were dilated and not reacting to light. MRI of head and neck showed a soft tissue mass lesion with epicenter in the right nasal cavity. Laterally, the tumor extended into the right maxillary antrum. Superiorly, it invaded into bilateral ethmoid sinuses and the sphenoid sinus. The lesion caused destruction of medial wall of orbit and extension into retro-orbital space with displacement of medial rectus and optic nerve. There was extracranial extension into the cavernous sinus with encasement of intracranial portion of the carotid artery. Posteriorly, there was extension into nasopharynx and pterygopalatine fossa (Figs 2A to D). Biopsy of the lesion was done and the histopathology picture showed normal pseudostratified columnar ciliated epithelial lining with normal underlying mucous glands, few fragments showed infiltration of stroma by cluster and cords of neoplastic cells having high nuclear cytoplasmic ratio, pleomorphic hyperchromatic nuclei with inconspicuous nucleoli and condensed chromatin. Prominent crushing artifacts and nuclear molding was prominent. Areas of necrosis and increased mitotic activity were also seen (Fig. 3). Immunohistochemical profile showed positivity for cytokeratin, chromogranin, synaptophysin and EMA. Supporting cells at the periphery did not stain for S-100. CD-45(LCA),CD99 and HMB45 were negative thereby ruling out lymphoma, primitive neuroectodermal tumor (PNET) and melanoma respectively (Figs 4A to D). The histological diagnosis, hence, was small cell neuroendocrine carcinoma of the sinonasal region. 18FDG-PET-CT scan showed the primary site to be limited to the right nasoethmoid complex with no other sites of uptake. CECT scan of chest and abdomen were normal. The patient was diagnosed as primary small cell neuroendocrine tumor stage cT4N0M0 (IVa). He was treated with induction chemotherapy with three cycle of injection Etoposide and

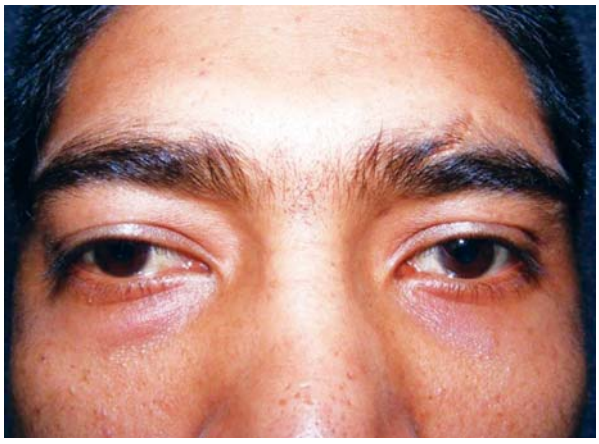
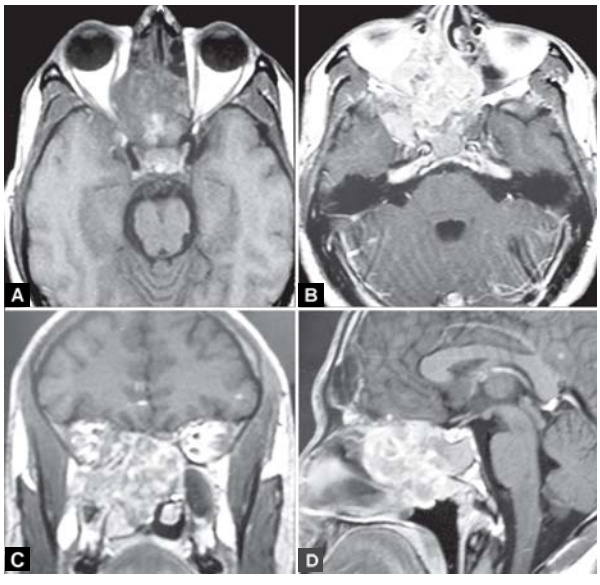


Fig. 1: Broadening of nasion



Figs 2A to D: MR Imaging (A and B) Cross-section view: Intraorbital extension with involvement of optic nerve on right side, (C) Coronal view: Intracranial extension, (D) Sagittal view: Sphenoid invasion

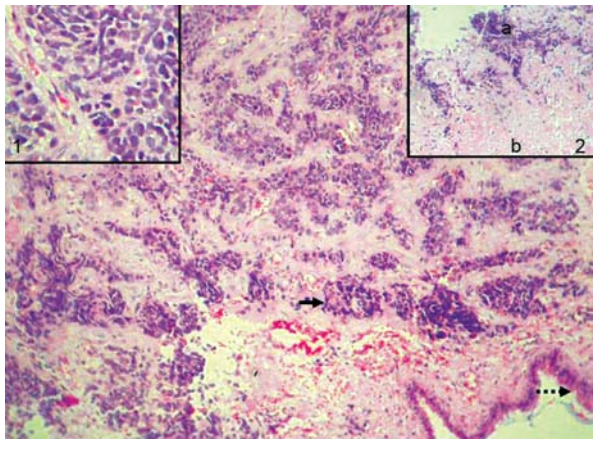
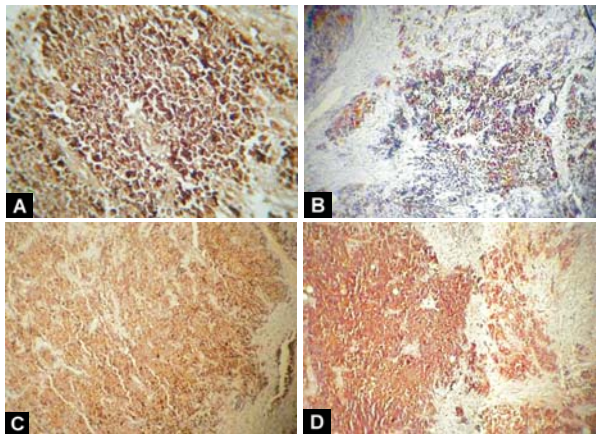


Fig. 3: H and E 10X: Nasal mucosa (broken arrow) with undifferentiated small cells underlying (solid arrow). Inset 1: H and E 40X: cells with high N:C ratio and granular cytoplasm. Inset 2: H and E 10X: crush artifacts (a) with necrosis (b)



Figs 4A to D: Immunohistochemistry profile: (A) Chromogranin, (B) cytokeratin, (C) synaptophysin, (D) EMA

Cisplatin followed by concomitant chemoradiotherapy (5940cGy/33/59 days) and three cycles of injection Etoposide and Cisplatin. Patient was re-evaluated 6 weeks after completion of therapy. There was clinical complete response. Patient has been followed up regularly on 3 months intervals with a combination of clinical examination and PET-CT. The patient has been disease free for last 20 months.

DISCUSSION

Small cell carcinoma (SmCC) is a histological subtype amongst a broad group of sinonasal malignancies, together known as sinonasal neuroendocrine tumors. Other subtypes include esthesioneuroblastoma (ENB), sinonasal undifferentiated carcinoma (SNUC) and neuroendocrine carcinoma (NEC).[1] SmCC, similar to oat-cell carcinoma of the lungs, is reported to arise in the nasal cavity and paranasal sinuses. As demonstrated in our case, the tumor is almost in an advanced stage by the time it comes to attention and this reflects its aggressive nature. Several sinuses are nearly always involved. The initial presentation of nasal obstruction, nasal discharge and recurrent epistaxis is practically indistinguishable from that of more benign diseases and hence is likely to result in delay in presentation.[2] Aggressive involvement of the right optic nerve and resultant diminished vision prompted a more thorough investigation in our case. This is similar to experience reported earlier.[3] SmCC has also been reported to be associated with multiple endocrine neoplasia,[4] though this was not our experience. SmCC has a morphologic, immunohistochemical and ultrastructural appearance similar to that of its pulmonary homolog.[5] The main differential diagnosis is olfactory neuroblastoma (esthesioneuroblastoma). While morphologically both have similar appearance, they share immunophenotype characteristic too. Presence of an organized rim of S-100 protein positive supporting cells (Schwann cells), synaptophysin positivity and CD 99 (MIC gene product)

negativity may help differentiate esthesioneuroblastoma from SmCC.[6,7] In our case, absence of S-100 positive supporting cells in the periphery with immunohistochemistry, suggestive of small cell neuroendocrine tumors, favored a diagnosis of SmCC over esthesioneuroblastoma.

As the tumor is rare, the treatment advised is controversial. While radiation therapy, with or without chemotherapy is generally advised,[7] chemotherapy and surgery have also been described.[8,9]

Thus, SmCC represents one rare histological subtype of neuroendocrine tumors affecting the sinonasal region. It is locally aggressive and, due to nonspecific features at initial presentation, it presents late. It is a histopathological challenge to differentiate it from esthesioneuroblastoma, which has a much more indolent course and better prognosis. A combination of histomorphology, immunohistochemical and ultrastructural characteristic needs to be considered to do so. Due to rarity of this condition, there is no consensus on its management guideline, but given its aggressive nature, it is perhaps prudent to treat it on lines of its pulmonary homolog with chemoradiation.

REFERENCES

1. Frazier SR, Kaplan PA, Loy TS. The pathology of extra pulmonary small cell carcinoma. Semin Oncol 2007;34:30-38.
2. Mendis D, Malik N. Sinonasal neuroendocrine carcinoma: A case report. Ear Nose Throat J 2008;87:280-82,293.
3. Avitia S, Osborne RF. Blindness: A sequela of sinonasal small cell neuroendocrine carcinoma. Ear Nose Throat J 2004;83: 530-32.
4. Westerveld GJ, van Diest PJ, van Nieuwkerk. EB Neuroendocrine carcinoma of the sphenoid sinus: A case report. Rhinology 2001;39:52-54.
5. Hatoum GF, Patton B, Takita C, et al. Small cell carcinoma of the head and neck: The university of Miami experience. Int J Radiat Oncol Biol Phys 2009;74:477-81.
6. Devaney K, Wenig BM, Abbondanzo SL. Olfactory neuroblastoma and other round cell lesions of the sinonasal region. Mod Pathol 1996;9:658-63.
7. Dulguerov P, Allal AS, Calcaterra TC. Esthesioneuroblastoma: A meta-analysis and review. Lancet Oncol 2001;2:683-90.
8. González-García R, Fernández-Rodríguez T, Naval-Gías L, et al. Small cell neuroendocrine carcinoma of the sinonasal region: A proposal of a case. Br J Oral Maxillofac Surg 2007;45: 676-78.
9. Babin E, Rouleau V, Vedrine PO. Small cell neuroendocrine carcinoma of the nasal cavity and paranasal sinuses. J Laryngol Otol 2006;120:289-97.