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Primary Sinonasal Neuroendocrine Carcinoma Invading the Orbit: Case Report and Review of Literature

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Abstract **Case Report**

Primary sinonasal neuroendocrine carcinoma (SNEC) is a rare aggressive sinonasal malignancy which typically occurs in the ethmoidal bone or sinuses, with a slight male preponderance. No risk factors have been identified. With a very few reported cases. Most patients present in advanced stages due to the lack of significant symptoms. Nomenclature has been ambiguous, but SNECs can be classified as well-, moderately- or poorly differentiated. Its treatment is not well established. We report the case of a large cell neuroendocrine carcinoma (LCNEC) in a 34 years old patient presented the occurrence of a right epistaxis. The beginning of the symptomatology goes back to 1 year before, with right nasal obstruction of progressive onset, associated with right epistaxis and bloody rhinorrhea with right chronic eye watering. Nasosinusal MRI shows a right nasal mass enhanced strongly after injection of gadolinium, extension to contralateral ethmoidal cells with orbital bone lysis and cribriform plate of the Ethmoid bone. The Histopathologic analysis of the mass biopsy showed a poorly differentiated carcinomatous tumor proliferation with positive neuroendocrine markers. The multidisciplinary decision taken was the treatment by chemotherapy before surgery.

Keywords: Tumor - neuroendocrine -carcinoma-sinonasal.

INTRODUCTION

Neuroendocrine carcinoma is relatively uncommon in the sinonasal region. Very few cases of sinonasal large cell neuroendocrine carcinoma have been reported till date. In recent years, this variant has been recognized as a distinct entity. Otherwise, no agreement for adequate management has been reached. These aggressive tumors are requiring multimodality treatment that includes surgery as initial treatment and chemoradiotherapy.

The focus of this paper is to report such extremely rare case and highlight recent developments in this pathology.

CASE REPORT

Mr F.B a 34-year-old male patient presented in our institution because of the occurrence of a right epistaxis. First of all, the patient is smoking and use cannabis.

The beginning of the symptomatology goes back to 1 year before, with right nasal obstruction of progressive onset, associated with right epistaxis and bloody rhinorrhea with right chronic eye watering.

The patient didn't present neurological signs nor cervical lymph nods. Nasal Endoscopy examination revealed burgeoning mass bleeding on contact and filling the right nostril vestibule.

Otherwise, ophthalmologic examination found right exophthalmia normal visual acuity and ocular mobility on both sides.

Sinonasal computed tomography (CT) found a right nasal process measuring 46X30X37 mm, enhanced after injection of contrast product, invading the right orbit with the left nasal fossae.

Otherwise, there is no extension involvement at the level of the cavum. Nasosinusal MRI shows a right nasal mass enhanced strongly after injection of gadolinium, extension to contralateral ethmoidal cells with orbital bone lysis and cribriform plate of the Ethmoid bone (Figure 1).

The Histopathologic analysis of the mass biopsy showed a poorly differentiated carcinomatous tumor proliferation reshaped and necrotic, with polypoid mucosa cells arranged in massive lobules rounded nuclei sparse cytoplasm. The immunohistochemical study of the tumor biopsy found positive pan cytokeratin and CK7 with positive neuroendocrine markers: Chromogranin A, synaptophysin and CD56, with a proliferation index Ki67 of 60%.

A review of the immunohistochemical analysis to look for neuroendocrine markers has been initiated and other neuroendocrine markers are not currently available in our institution.

The multidisciplinary decision taken was the treatment by chemotherapy before surgery.

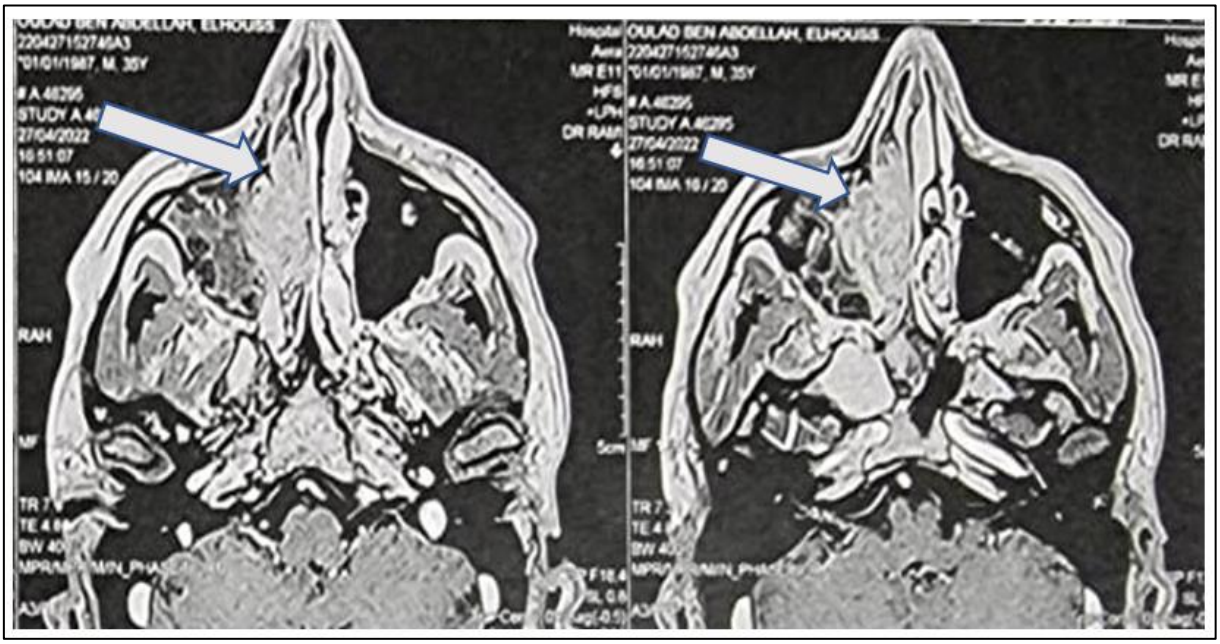

Figure 1: Axial view of MRI head and orbits with contrast demonstrating the right orbital tumor (white arrows) displacing the globe anterolaterally and extending to the paranasal sinuses and left nasal cavity

DISCUSSION

Primary sinonasal neuroendocrine carcinomas (SNECs) are rare and account for 3 to 5% of all sinonasal malignancies [1-3].

SNECs are categorized by their differentiation grade into well-, moderately- and poorly differentiated. The latter group is further subdivided into small and large cell neuroendocrine carcinoma. Poorly differentiated large cell SNEC are in fact classified as 'sinonasal undifferentiated carcinoma' (SNUC), whereas poorly differentiated small cell SNEC is referred to as 'sinonasal small cell carcinoma'.

Importantly, differentiation grade determines prognosis rather than TNM staging. Well-to moderately differentiated neuroendocrine carcinomas are associated with lower metastatic rates and better survival (5-year disease-specific survival approximatively 70%) compared to the rarer poorly differentiated neuroendocrine carcinomas (5-year approximatively 40%) [4-7].

Most patients present at advanced stages due to the lack of significant symptoms. Advanced tumors may invade the skull, orbit or brain. Ophthalmic manifestations include exophthalmos, reduced vision and restriction in ocular motility. Ectopic hormone secretion has been described in a handful of cases.

Staging is of limited value in predicting prognosis and recent literature clearly highlights the importance of histological diagnosis, particularly differentiation grade, in determining the prognosis and predicting treatment response [4-7].

Ectopic hormone secretion has been described in a handful of cases. These cases concerned patients with SNEC or SmCC with elevated levels of ACTH, beta-MSH, calcitonin, serotonin or ADH [1].

Differential diagnoses include sinonasal undifferentiated carcinoma (SNUC), malignant melanoma, and lymphoma. Rarity of cases and overlapping morphologic and immunohistochemical features pose a conceptual challenge in the diagnosis of Large Cell Neuroendocrine Carcinoma LCNEC and its differentiation from SNUC and high-grade olfactory neuroblastoma (ONB) [1].

Aggressive trimodality therapy seems to be the most effective approach, although survival remains poor. Surgery supplemented with postoperative radiotherapy or concomitant chemoradiotherapy is accepted as the primary treatment for localized disease. For patients with systemic disease, palliative chemotherapy or best supportive care remains appropriate options [8].

In a meta-analysis, van der Laan *et al*., rapported that, irrespective of the histological diagnosis of NEC, surgery has a beneficial effect on survival and should be the cornerstone of any treatment strategy. This is supported by both the univariate and multivariate analyses in which treatment (combinations) incorporating surgery produced the best results. Otherwise, Radiotherapy as monotherapy should not be performed in curative setting.

No benefit from the application of chemotherapy could be deduced from his results. Chemotherapy as monotherapy had the worst 5-year median disease-specific survival, with no patients surviving regardless of tumor subtype. However, no strong recommendations can be made in this regard, due to semantic deficiencies in the literature [1].

We concluded that neuroendocrine carcinoma of the nasal cavity is an extremely rare and destructive neoplasm. Clinical presentations of this uncommon malignancy include non-specific symptoms. The improvement in treatment outcome over time is best explained by the shift towards multimodality therapy as advocated by several authors.

Compliance with ethical standards
Disclosure of conflict of interest: None

Statement of Informed Consent: Informed consent was obtained from all individual participants included in the study

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