World Journal of Advanced Research and Reviews
eISSN: 2581-9615 CODEN (USA): WJARAI
Cross Ref DOI: 10.30574/wjarr
Journal homepage: https://wjarr.com/

(CASE REPORT)

Neuroendocrine carcinoma of the nasal cavity and paranasal sinuses: A case report and literature review

Driss Elidrissi [1,*], Omar Oulghoul [1], Omayma bourht [1], Mohamed Chehbouni [1], Youssef Lakhdar [1], Othmane Benhoummad [2], Youssef Rochdi [1] and Abdelaziz Raji [1]

[1] *Department of ENT and HNS Surgery, Mohammed VI University Hospital, Marrakech, Morocco.*
[2] *Department of ENT and HNS Surgery, Faculty of Medicine and Pharmacy of Agadir, Ibn Zohr University, Agadir, Morocco.*

World Journal of Advanced Research and Reviews, 2024, 24(02), 2259–2264

Publication history: Received on 11 October 2024; revised on 22 November 2024; accepted on 25 November 2024

Article DOI: https://doi.org/10.30574/wjarr.2024.24.2.3553

Abstract

Neuroendocrine tumors are defined as epithelial neoplasms with predominant neuroendocrine differentiation. They can occur in almost any organ of the body. The nasal cavity and paranasal sinuses are a rare site of neuroendocrine carcinoma. Recurrences are frequent, and prognosis is poor. The difficulty of pathological diagnosis and the rarity of this malignancy have hampered progress in understanding the clinical course and improving outcomes. We report a rare case of a neuroendocrine tumor of the nasal cavity and ethmoidal sinuses with invasion of the orbit in a 35-year-old male patient. The literature is reviewed and aetiopathogenic theories, diagnostic procedures and surgical approaches are discussed.

Keywords: Neuroendocrine Carcinoma; Nasal cavity; Paranasal sinuses; Endoscopic surgery; Radiotherapy

1. Introduction

In head and neck region, neuroendocrine carcinoma (NEC) is most frequently found in the larynx, followed by salivary glands [1]. It is very rarely found in nasal cavity and paranasal sinuses, accounting for around 5% of malignancies at these sites [2] [3]. The World Health Organization (WHO) classifies NEC into three subtypes: typical carcinoid (well-differentiated); atypical carcinoid (moderately differentiated); and small-cell carcinoma (poorly differentiated) [4].

However, understanding the pathogenesis of this disease has been elusive due to its rarity and the complexity of histological diagnosis.

Due to the rarity of nasosinusal neuroendocrine carcinomas, no agreement on appropriate management has been reached.

The aim of the current study is to report a descriptive study and analyze the available information relatively important of a rare tumor.

2. Case report

A 35-year-old chronic smoker consulted us with a right unilateral nasal obstruction evolving for 1 year, associated with recurrent homolateral epistaxis of low abundance, complicated 6 months later by homolateral exophthalmos with no other associated signs.

* Corresponding author: Driss Elidrissi

Clinical examination revealed a right paranasal swelling firm, associated with irreducible, non-pulsatile right exophthalmos (figure 1).

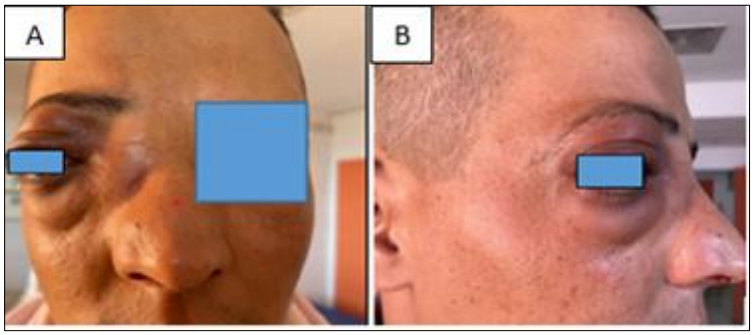

Figure 1 Front (A) and profile (B) image showing exophthalmos of the right eye with homolateral paranasal swelling

Nasal endoscopy showed a budding mass filling the entire right nasal cavity, with purulent secretions. Preoperative ophthalmological examination revealed exophthalmos with no decrease in visual acuity.

Computed tomography (CT) revealed a lesional process centered on the ethmoidal cells and the right nasal fossa, spontaneously isodense, intensely and heterogeneously enhancing after injection of contrast, measuring 46 x 30 x 37 mm, responsible for infiltration of extra conical internal fat of the right orbit and coming into contact with homolateral internal muscle, with lysis of upper nasal septum (Figure 2).

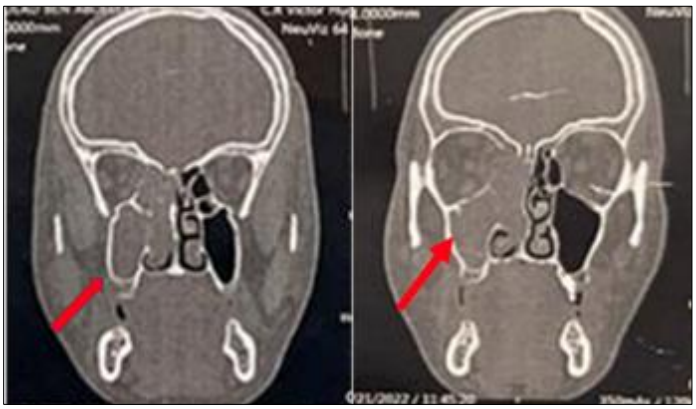

Figure 2 Coronal CT image showing a lesional process occupying the ethmoidal cells and right nasal fossa with extension to the homolateral orbit

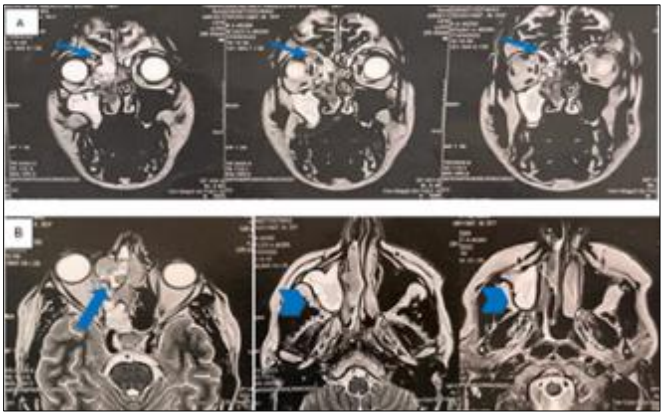

Figure 3 A- Coronal MRI showing a heterogeneous lesional process of the right nasal fossa with lysis of the right orbital wall. B- Axial MRI showing extension of the tumor to the contralateral ethmoidal cells, with retention of the right maxillary sinus

Magnetic resonance imaging (MRI) found an aggressive, heterogeneous lesion in the right nasal cavity measuring 31 x 42 mm, extending into the contralateral ethmoidal cells and causing lysis of the right orbital wall and ethmoidal cribriform plate, with invasion of the extra conical fat of the right orbit and without involvement of the orbital muscles (Figure 3).

A biopsy was performed. Histological analysis was in favor of a poorly differentiated tumor proliferation of probable carcinomatous origin, extensively remodeled and necrotic.

Immunohistochemical (IHC) study revealed that the neoplastic cells were negative for S-100, and positive for cytokeratin, synaptophysin and chromogranin. Immunohistochemical appearance consistent with poorly differentiated neuroendocrine carcinoma (Figure 4).

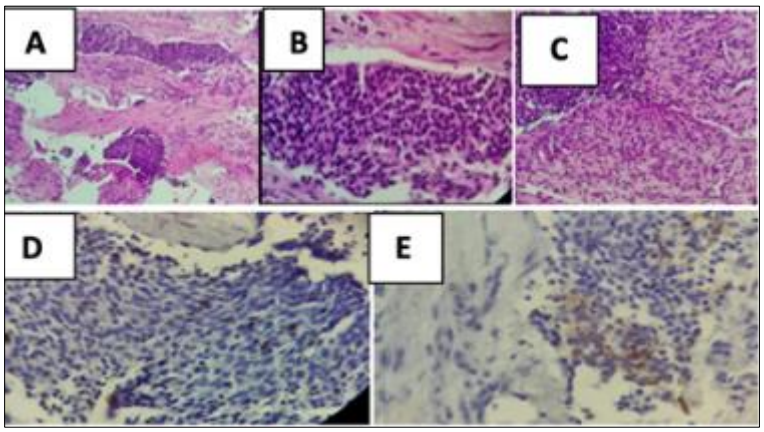

Figure 4 A: slide with Hematoxylin and Eosin (HE) staining, showing fibrous tissue with infiltrating tumor proliferation arranged in clumps. B+C: 2 high-magnification HE slides showing small, contiguous tumor cells with ovoid hyperchromatic nuclei with irregular outlines in places, cytoplasm is sparse. D: Immunohistochemical slide showing focal and moderate nuclear expression of anti-p63 Ac in tumor cells. E: Immunohistochemical slide showing focal cytoplasmic expression of anti-chromogranin ac in tumor cells

A work-up for extension and multiple endocrine neoplasia was requested in this context and proved negative.

This case was discussed in a multidisciplinary staff meeting with ophthalmologists and oncologists. We opted for endoscopic surgery with exenteration of the right eye. When the patient refused surgery, he was referred to oncology for induction chemotherapy (Cisplatin and Etoposide) in 3 cycles. A follow-up facial MRI revealed the persistence with slight regression of the lesion process in the right nasal cavity, as well as regression of the right orbital infiltration (Figure 5).

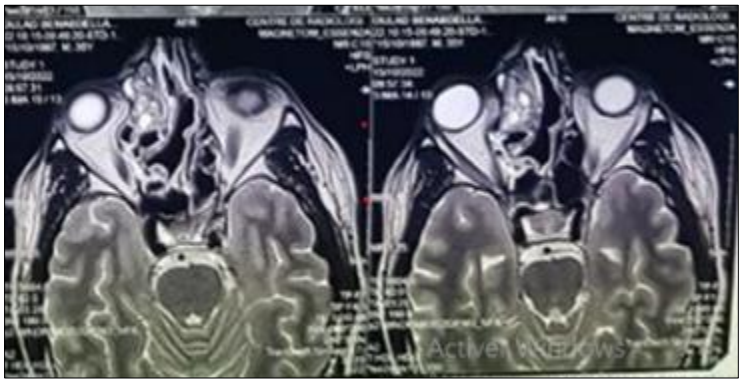

Figure 5 T2 axial section facial MRI showing persistent lesion process in the right nasal fossa, with regression of the right orbital infiltration

Chemotherapy was discontinued, and surgery was reconsidered after discussion with the ophthalmic surgeons. We opted for a total excision of the endonasal mass (Figure 6, 7) with recuts at the level of the papyraceous lamina and preservation of the orbit, followed by adjuvant radiotherapy.

Histological analysis of the surgical specimen confirmed the diagnosis revealing a neuroendocrine carcinoma, with lesion-free resection margins.

After 10 months of follow-up, no distant organ metastasis or recurrence in the primary tumor region was detected. The short-term treatment was effective, although the follow-up is only 10 months, long-term follow-up work still needs to be continued.

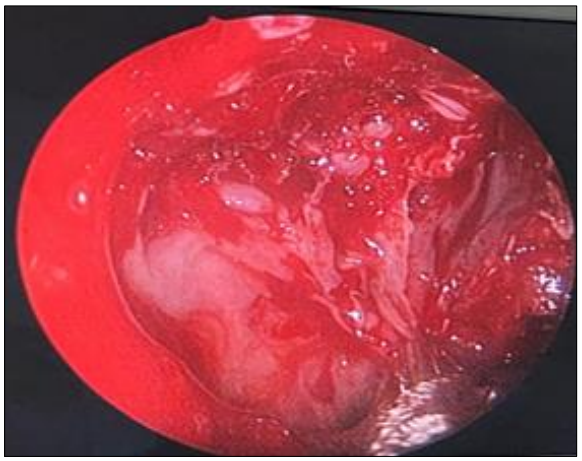

Figure 6 Endoscopic intraoperative view of tumor

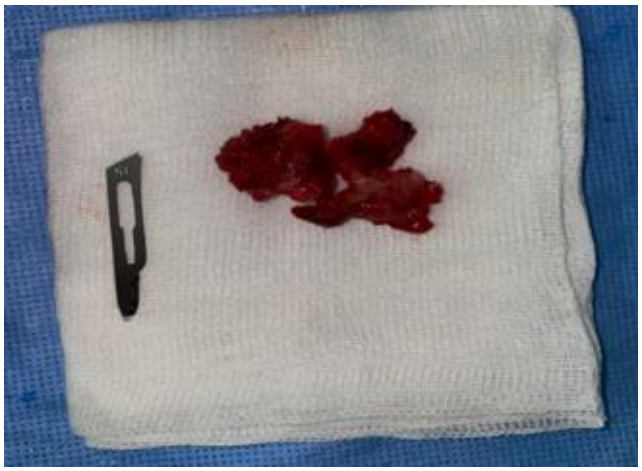

Figure 7 Surgical excision piece

3. Discussion

Neuroendocrine carcinoma occurs mainly in lungs, accounting for around 20% of primary lung carcinomas. Extra-lung localizations of these carcinomas account for 4% of all cases [5], and a limited number of cases of localization in nasal and paranasal cavities have already been reported.

Neuroendocrine carcinoma of naso-sinus tract occurs more frequently in men in the 5th and 6th decades. The ethmoidal sinuses are the most affected [6, 7]. A history of smoking was common in patients with tumors of the head and neck region. A strong association with smoking was identified in cases of pulmonary neuroendocrine carcinomas. However,

a strong association with smoking has not been identified in neuroendocrine carcinomas of the paranasal sinuses to date. No specific etiological factors have been identified [6, 7].

The clinical features of nasosinusal neuroendocrine carcinoma are non-specific and similar to those of other nasosinusal tumors. Common presentations include nasal obstruction, epistaxis, a facial mass and/or facial pain. The majority of patients have advanced disease at presentation [8]. Extensive involvement including the skull, orbit and brain may be observed. Ophthalmic manifestations include exophthalmos, impaired visual acuity and limited ocular mobility. Local pain, anosmia and metastatic cervical lymph nodes have also been described [9].

A review of the published literature reveals that these tumors are recurrent and locally destructive.

Anatomical imaging can reveal signs of malignancy, such as the existence of a lytic process. Magnetic resonance imaging with gadolinium injection improves differentiation between an inflammatory reaction, a tumor and/or fluid retention. It also identifies the anatomical relationship between the tumor and the meninges.

The treatment of nasosinusal neuroendocrine carcinomas has not been systematically evaluated, due to the small number of cases. No agreement on appropriate management has been reached, and some recommendations have been based on retrospective data. Surgery, radiotherapy and chemotherapy alone or in combination have been used [4].

In the 1980s, surgery followed by radiotherapy was the routine approach to treating neuroendocrine tumors. In the late 1990s, Fitzek et al [10] and Bhattacharyya et al [11] showed promising results with induction chemotherapy with cisplatin and etoposide followed by radiotherapy in the treatment of these tumors. A spectacular response was achieved even in the case of large or unresectable tumors. Although Bhattacharyya et al [10], Fitzek et al [11], and Babin et al [8] have proposed chemotherapy followed by radiotherapy with surgery reserved for non-responders as a treatment protocol. A few recent studies have shown that surgery as initial treatment followed by postoperative chemoradiotherapy is associated with better disease control and overall survival in the treatment of nasosinusal neuroendocrine carcinomas, even in poorly differentiated small-cell neuroendocrine carcinomas.

The prognosis of these tumors is very poor, due to the high rate of metastasis observed. Common metastatic sites are brain, lung, bone and skin. Non-lung neuroendocrine carcinomas are associated with a survival rate of up to 13% at five years, however [8].

4. Conclusion

Neuroendocrine carcinoma of the nasal cavity is a rare, highly aggressive tumor with a high incidence of local recurrence and early distant metastases. Further studies are needed to assess optimal management and develop standardized treatment protocols.

Compliance with ethical standards

Disclosure of conflict of interest

There are no conflicts of interest to declare related to this research.

Statement of ethical approval

While formal ethical approval was not obtained for this study, we ensured that all aspects of the research were conducted ethically and with respect for the rights and well-being of the participants.

Statement of informed consent

Informed consent was obtained from all participants involved in the study, and this information has been appropriately included in the manuscript.

References

[1] Renner G. Small cell carcinoma of the head and neck: a review. Semin Oncol. 2007;34(1):3-14.PubMed | Google Scholar

[2] Patel TD, Vazquez A, Dubal PM, Baredes S, Liu JK, Eloy JA. Sinonasal neuroendocrine carcinoma: a population-based analysis of incidence and survival. Int Forum Allergy Rhinol. 2015;5

[3] Qin Zhao, Jinlong Wei, Chengbin Zhang, Lingbin Meng, Bin Wang, Ling Gao1,Xin Jiang,Large-cell neuroendocrine carcinoma of nasal cavity and paranasal sinuses after successful curative therapy: a case report and literature review. 2019

[4] Nagesh T. Sirsath,1 K. Govind Babu,1 Umesh Das,1 and C. S. Premlatha2 Paranasal Sinus Neuroendocrine Carcinoma: A Case Report and Review of the Literature , 2013

[5] Bell D, Hanna EY. Sinonasal undifferentiated carcinoma: morphological heterogeneity, diagnosis, management and biological markers. Expert Rev Anticancer Ther 2013;13:285–296.

[6] E. Babin, V. Rouleau, P. O. Vedrine et al., "Small cell neuroendocrine carcinoma of the nasal cavity and paranasal sinuses," *The Journal of Laryngology & Otology*, vol. 120, no. 4, pp. 289– 297, 2006.

[7] G. Han, Z. Wang, X. Guo, M. Wang, H. Wu, and D. Liu, "Extrapulmonary SNEC of paranasal sinuses," *Journal of Oral and Maxillofacial Surgery*, vol. 70, no. 10, pp. 2347–2351, 2012.

[8] E. H. Mitchell, A. Diaz, T. Yilmaz et al., "Multimodality treatment for sinonasal neuroendocrine carcinoma," *Head & Neck*, vol. 10, pp. 1372–1376, 2012.

[9] E. G. Silva, J. J. Butler, B. Mackay, and H. Goepfert, "Neuroblastomas and neuroendocrine carcinomas of the nasal cavity: a proposed new classification," *Cancer*, vol. 50, no. 11, pp. 2388– 2405, 1982.

[10] M. M. Fitzek, A. F. Thornton, M. Varvares et al., "Neuroendocrine tumors of the sinonasal tract: results of a prospective study incorporating chemotherapy, surgery, and combined proton-photon radiotherapy," *Cancer*, vol. 94, no. 10, pp. 2623– 2634, 2002.

[11] N. Bhattacharyya, A. F. Thornton, M. P. Joseph, M. L. Goodman, and P. C. Amrein, "Successful treatment of esthesioneuroblastoma and neuroendocrine carcinoma with combined chemotherapy and proton radiation: results in 9 cases," *Archives of Otolaryngology*, vol. 123, no. 1, pp. 34–40, 1997.