Journal of
Anatomy and Physiological Studies

CASE REPORT

A Case Report: Esthesioneuroblastoma as a Cause of Nasal Obstruction

Esteban Espinoza-García[1]*, Mario Álvarez-Tapia[2], Nicolás Meza-Saavedra[3]

[1]Neurosurgeon, San Camilo Hospital, Chile
[2]ENT Specialist, San Camilo Hospital, Chile
[3]School of Medicine, Universidad de Valparaíso, San Felipe, Chile

Abstract

Esthesioneuroblastoma, also called olfactory neuroblastoma, is a rare malignant neoplasm of the nasal cavity. It corresponds to 0.3% of upper airway tumors and about 3-6% of all intranasal tumors. Clinical presentation of esthesioneuroblastoma is usually quite nonspecific, with nasal obstruction and recurrent epistaxis being the most common symptoms. Other symptoms include headache, facial pain, sinusitis and anosmia. Because the early symptoms are quite nonspecific, the diagnosis is usually made late, so a high index of suspicion becomes crucial to improve the early detection of this type of tumor. We present a case of 63-year-old patient, who consults in the ENT department for multidirectional nystagmus and nasal obstruction of 6 months of evolution. A study is performed with CT and MRI, with extensive osteolytic skull base lesion appearing. A biopsy was performed using nasofibroscopy, confirming a grade II esthesioneuroblastoma. This tumor is a rare cause of nasal obstruction, but it must be included in differential diagnoses.

Keywords: Esthesioneuroblastoma, Olfactory Neuroblastoma, Case Report, Nasal Obstruction, Tumor

Introduction

Esthesioneuroblastoma (ENB) corresponds to an uncommon malignant neoplasm, originated from the olfactory neuroepithelium located in the upper portion of the nasal cavity that commonly invades both the cranial vault and the eye socket [1].

ENB has a bimodal distribution, without a clear predisposition for sex, with presentation peaks during the second and sixth decades of life [2, 3]. Its incidence is approximately 0.4 cases per million of population [2, 4, 5] and corresponds to 3 - 6% of all intranasal tumors [6]. ENB histogenesis was debate of subject during its first years of discovery, giving this tumor a variety of names across the years [7]. It was in 1960 that it was demonstrated that the tumor's origin was found in the neuroectoderm and olfactory epithelium. Hyams [8] proposed a histopathological classification system for the ENB, based on architecture, nuclear polymorphism, necrosis and rosette formation, accomplishing to establish an independent prognostic indicator [9].

The clinical presentation for the ENB is usually quite nonspecific, with nasal obstruction [10-12] being its main symptom, followed by epistaxis [10, 11]. Other symptoms include headaches, facial pain, sinusitis, anosmia and visual disturbances [13]. Because the early symptoms are quite nonspecific and can simulate benign processes, diagnosis of this tumor is usually late, with a delay of 6 - 12 months [10, 11] since symptoms onset until the confirmation of the diagnosis, hence a high suspicion rate it is fundamental to detect this tumor in early stages.

In 1967 Kadish [14] developed a staging system based on radiological features considering tumoral extension, classifying the ENB in 4 possible stages. This staging system was demonstrated to be a good prognostic indicator [15].

Case Details

We present a female patient of 63 years old, no previous history of diseases. The woman requested a first consultation with the ENT service, due to nasal obstruction and diplopia of 6 months evolution. Other clinical findings were not investigated at the physical examination, and the patient also referred to the presence of headache associated with insomnia and mood disorders.

A brain CT-scan was performed, which evidences extensive osteolytic skull base lesion (Figure 1). Subsequently, a nasofibroscopy was performed, accessing the right nostril, observing a pulsatile and vascularized mass infiltrating middle turbinate and occluding the choanae. The same lesion can be seen on the left nostril. A biopsy was performed, indicating the

***Correspondence to:** Esteban Espinoza-García, San Camilo Hospital, Salinas 1392, Mailbox 150, San Felipe, Chile, Tel: +56 9 342493200, Email: esteban[DOT]espinoza[AT]postgrado[DOT]uv[DOT]cl

Received: June 30, 2020; **Accepted:** July 09, 2020; **Published:** July 13, 2020

*This article is reviewed by "Sharadkumar Pralhad Sawant" (India)

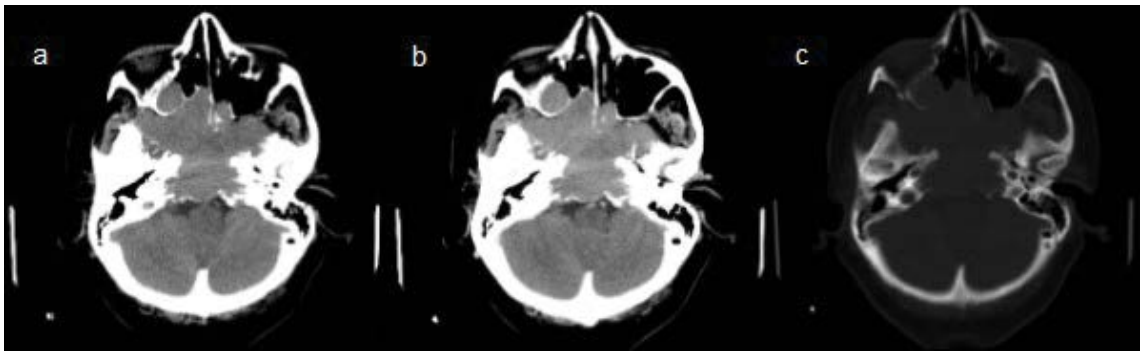

Figure 1: Brain CT; a & b: Without and with contrast respectively, showing an extensive isodense infiltrative lesion with low heterogeneous contrast enhancement; **c:** Bone window, observing extensive osteolytic skull base lesion.

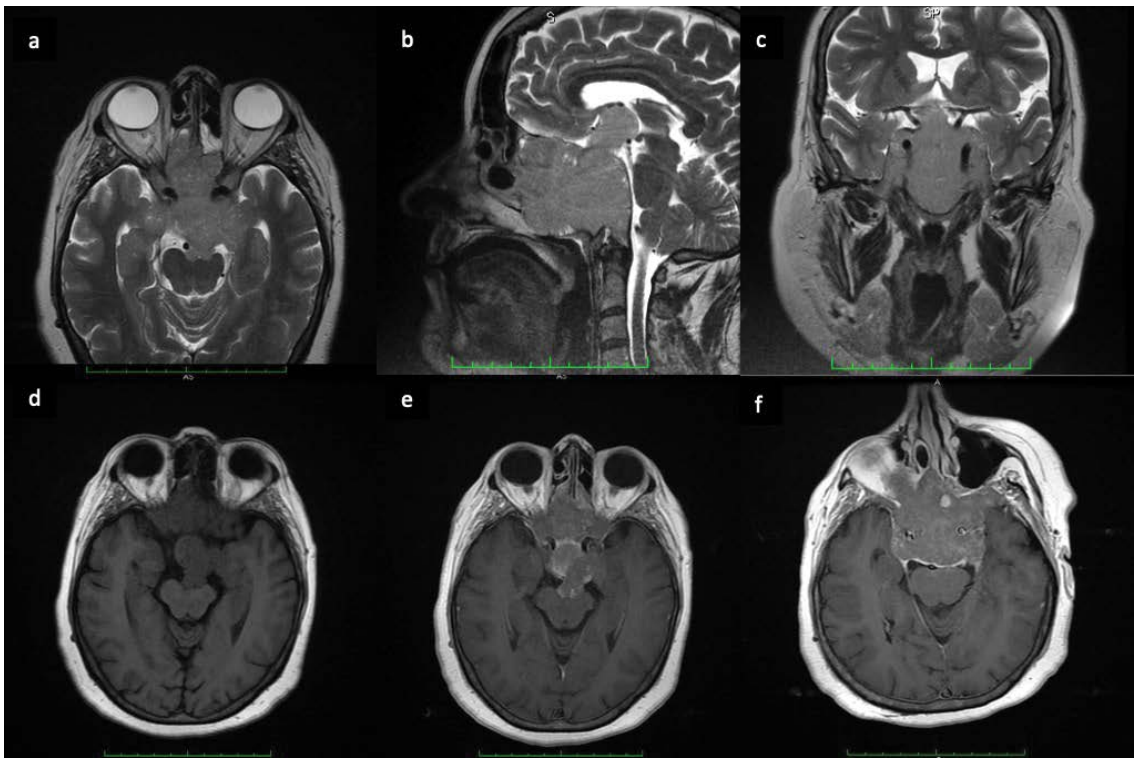

Figure 2: Brain MRI; a, b, c: T2 sequences, showing an extensive infiltrative lesion; **d, e:** T1 sequences observing a heterogeneous contrast enhancement; **f:** The infiltrative lesion enveloping both carotid siphons.

presence of a small cell tumor with positive neuroendocrine markers, compatible with a grade II ENB.

At the ophthalmological examination, a superior bitemporal quadrantopsia was found. In blood tests, a prolactin of 32.6 and a TSH of 5.45 were remarkable. Subsequently, an MRI was performed, which showed an extensive lesion of approximately 7 cm in larger diameter, which invades the sellar region, enveloping both carotid siphons and invading interpeduncular fossa, contacting in its inferior portion to the anterior arch of the atlas, and in its anterior region deforming turbinates and invading right maxillary sinus. This lesion is observed heterogeneous in T1 and T2 sequences, with heterogeneous and moderate contrast enhancement (Figure 2).

Discussion

ENB is a rare neuroectodermal tumor, originating in the olfactory epithelium and derived from neural crest cells, and accounts for 3-6% of all malignant tumors of the paranasal sinuses [6, 16]. Clinically, olfactory neuroblastoma manifests as nasal obstruction or epistaxis. It can show indolent behavior, promote local invasion, and generate distant metastases. It tends to invade the paranasal sinuses, orbits, and anterior cranial fossa. The most common metastases are to the lymph nodes of the neck, lungs, liver, and bone, such dissemination at the time of diagnosis being the main predictor of survival. Although there is no universally accepted staging system, the Kadish classification system is widely used. Imaging plays a key role in the accurate staging of ENB including both CT and MRI [17].

ENB does not have specific CT characteristics, presenting initially as a homogeneous soft tissue mass in the nasal vault, however, CT is essential for evaluation of the osseous involvement of the cribriform plate, fovea ethmoidalis, and lamina papyracea. The mass shows moderate and uniform enhancement. Scattered speckled calcifications are occasionally present. CT is also useful to assess regional neck and distant metastasis. MRI is superior in defining the soft tissue extent and offers more accurate assessment of suspected intracranial, orbital, or skull base invasion. ENB is usually hypointense to gray matter on T1-weighted images and intermediate to hyperintense on T2-weighted sequences. The tumor demonstrates homogeneous enhancement except for areas of necrosis or hemorrhage. The presence of intracranial cysts is highly suggestive of ENB [18, 19].

ENB is a rare disease should be considered as a possible cause of nasal obstruction when other more frequent causes are discarded, and suspicion is crucial at the time of the diagnosis.

Acknowledgments

None

Conflicts of interest

The authors declare that there is no conflict of interest regarding the publication of this article.

References

1. Toader C, Stroi M, Kozma A, Arsene DE (2018) Esthesioneuroblastoma: the complete picture – case report and review of the literature. *Rom J Morphol Embryol* 59:1211–1218. [View Article]
2. Fiani B, Quadri SA, Cathel A, Farooqui M, Ramachandran A, et al. (2019) Esthesioneuroblastoma: A Comprehensive Review of Diagnosis, Management, and Current Treatment Options. *World Neurosurg* 126:194–211. [View Article]
3. Gondim J, Ramos Jr J, Azevedo J, Carrero Jr FP, Tella Jr OI. (2002) Esthesioneuroblastoma: case report. *Arq Neuropsiquiatr* 60: 303–307. [View Article]
4. Uslu GH, Canyilmaz E, Zengin AY, Mungan S, Yoney A, et al. (2015) Olfactory neuroblastoma: A case report. *Oncol Lett* 10: 3651–3654. [View Article]
5. Thompson LDR (2009) Olfactory Neuroblastoma. *Head and Neck Pathol* 3: 252–259. [View Article]
6. Sepúlveda I, Herrera A, Delgado C, Platin E, Rosel P (2012) Esthesioneuroblastoma: a case report and review of the literature. *Int J Odontostomat* 6: 285–289. [View Article]
7. Oskouian RJ Jr, Jane JA Sr, Dumont AS, Sheehan JM, Laurent JJ, et al. (2002) Esthesioneuroblastoma: clinical presentation, radiological, and pathological features, treatment, review of the literature, and the University of Virginia experience. *Neurosurg Focus* 12: e4. [View Article]
8. Hyams VJ, Batsakis JG, Michaels L (1988) Atlas of Tumour Pathology. Tumors of the upper respiratory tract. Washington, DC: Armed Forces Institute of Pathology.
9. Rimmer J, Lund VJ, Beale T, Wei WI, Howard D (2014) Olfactory Neuroblastoma: A 35–Year Experience and Suggested Follow–up Protocol. *Laryngoscope* 124: 1542–9. [View Article]
10. Abdelmeguid AS (2018) Olfactory Neuroblastoma. *Curr Oncol Rep* 20: 7. [View Article]
11. Ow TJ, Bell D, Kupferman ME, Demonte F, Hanna EY (2013) Esthesioneuroblastoma. *Neurosurg Clin N Am* 24: 51–65. [View Article]
12. Benet S, Garabal JA, Luis J, Tacoronte L, Artazkoz JJ. (2017) Estesioneuroblastoma o neuroblastoma olfatorio. *Rev Otorrinolaringol Cir Cabeza Cuello* 77: 63–68. [View Article]
13. Su SY, Bell D, Hanna EY. (2014) Esthesioneuroblastoma, Neuroendocrine Carcinoma, and Sinonasal Undifferentiated Carcinoma: Differentiation in Diagnosis and Treatment. *Int Arch Otorhinolaryngol* 18: S149–S156. [View Article]
14. Kadish S, Goodman M, Wang CC (1976) Olfactory neuroblastoma. A clinical analysis of 17 cases. Cancer 37: 1571–1576. [View Article]
15. König M, Osnes T, Jebsen P, Evensen JF, Meling TR. (2018) Olfactory neuroblastoma: a single–center experience. *Neurosurg Rev* 41: 323–331. [View Article]
16. Ferreira MCF, Tonoli C, Varoni ACC, Gusmon CC, Alvarenga M, et al. (2007) Estesioneuroblastoma. *Rev Ciênc Méd* 16: 193–198. [View Article]
17. Naves AA, da Silveira Filho LG, Etchebehere R, Ribeiro Júnior HA, Lima Junior FVA (2017) Esthesioneuroblastoma. *Radiol Bras* 50: 341–342. [View Article]
18. Dublin AB, Bobinski M (2016) Imaging Characteristics of Olfactory Neuroblastoma (Esthesioneuroblastoma). *J Neurol Surg B Skull Base* 77: 1–5. [View Article]
19. Som PM, Lidov M, Brandwein M, Catalano P, Biller HF (1994) Sinonasal esthesioneuroblastoma with intracranial extension: marginal tumor cysts as a diagnostic MR finding. *AJNR Am J Neuroradiol* 15: 1259–1262. [View Article]

Citation: Espinoza-García E, Álvarez-Tapia M, Meza-Saavedra N (2020) A Case Report: Esthesioneuroblastoma as a Cause of Nasal Obstruction. J Anat Physiol Stud 3: 001-003.