Scholars Journal of Medical Case Reports

OPEN ACCESS

Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online)
Journal homepage: https://saspublishers.com

Agressive Esthesioneuroblastoma about A Case

Ihssane Allouch[1*], Hanaa Rahim[1], Najour Belhaj[1], Razika Bencheikh[1], Mohamed Anas Benbouzid[1], Leila Essakalli[1]

[1]Service ORL et CCF de l'hôpital des spécialités de Rabat, CHU Rabat Salé

DOI: 10.36347/sjmcr.2021.v09i05.003 | **Received:** 17.03.2021 | **Accepted:** 24.04.2021 | **Published:** 07.05.2021

***Corresponding author:** Ihssane Allouch

Abstract **Case Report**

Introduction: Esthesioneuroblastomas (ENB) or olfactory neuroblastomas are rare tumors derived from the olfactory epithelium. These tumors have a propensity for local invasion into surrounding structures and distant metastasis, most commonly to the neck, lungs, and bones. We present a case of a 12-year-old child with olfactory neuroblastoma with lymph node metastasis with early recurrence. ***Case Report:*** It is a 12 year old child, without pathological history who presented an evolving right nasal obstruction for 8 months, permanent, associated with a rhinorrhea and intermittent epistaxis, associated with a mass of the ipsilatéral parotid and the submandibular region. The patient underwent a nasosinnus and cervical scan, a biopsy under general anesthesia, an anatomopathological study with immunohistochemistry which were in favor of esthesioneuroblastoma. ***Conclusion***: ENB is a rare malignant tumor of the nasal cavity. It is characterized by clinical polymorphism and difficulty in histological diagnosis. CT and MRI are one of the therapeutic strategy keys and then to post-treatment monitoring.

Keywords: Esthesioneuroblastoma, lymph node metastasis, surgery.

INTRODUCTION

Ethesioneuroblastoma (ENB), or olfactory neuroblastoma, is a rare malignant tumor arising from the olfactory sensory neuroepithelium, which accounts for 2%e6% of all primary sino-nasal malignancies [1, 2]. ENB was first described by Berger and Luc in 1924, No risk factor is currently recognized for this type of tumor, ENB has a bimodal distribution with no gender predilection and tends to occur around the second and sixth decade of life; however, sporadic cases have been reported in children younger than 10 years [3-5].

The natural history of ENB is varied and heterogenous, with some tumors demonstrating annindolent course and others exhibiting a highly aggressive behavior characterized by local recurrences and/or distant metastasis [3].

We present a case of a 12-year-old child with olfactory neuroblastoma with lymph node metastasis with early recurrence.

CASE REPORT

It is a 12 year old child, without pathological history who presented an evolving right nasal obstruction for 8 months, permanent, associated with a rhinorrhea and intermittent epistaxis, associated with a mass of the ipsilatéral parotid and submandibular region. The clinical examination, showed a gray and pink mass occupying the entire right nasal and invaded the nasal septum and the contralateral nasal fossa, with a retro angulo maxillary mass extended to the parotid region and jugulo carotidien, hard, painful and fixed at the deep level (Figure-1).

A cervical and nasosinnusian CT scan showed a tumor occupying the two nasal cavities the nasopharynx the ethmoid sinus and lysing the posterior internal wall of the right maxillary sinus and the sphenoid sinus body with invasion of the latter two the scanner also showed the presence of cervical polyadenopathies on the right side (Figure-2).

The biopsy was performed under general anesthesia, the pathological and immunohistochemistry examination were in favor of an esthesio neuroblastoma , given the significant tumor extension, the decision was to do a chemotherapy at first, the post-chemotherapy scan showed tumor progression (Figure-3).

RESULTS

The child then underwent transfacial surgery with parotid lymph node dissection and groups Ib, II, III et V right (Figure-4), he was then referred for radiotherapy. Three weeks after the operative procedure

the child presented a recurrence of tumor at the level of the operated lymph node sites and at the level of the contralateral side, the child died during the assessment.

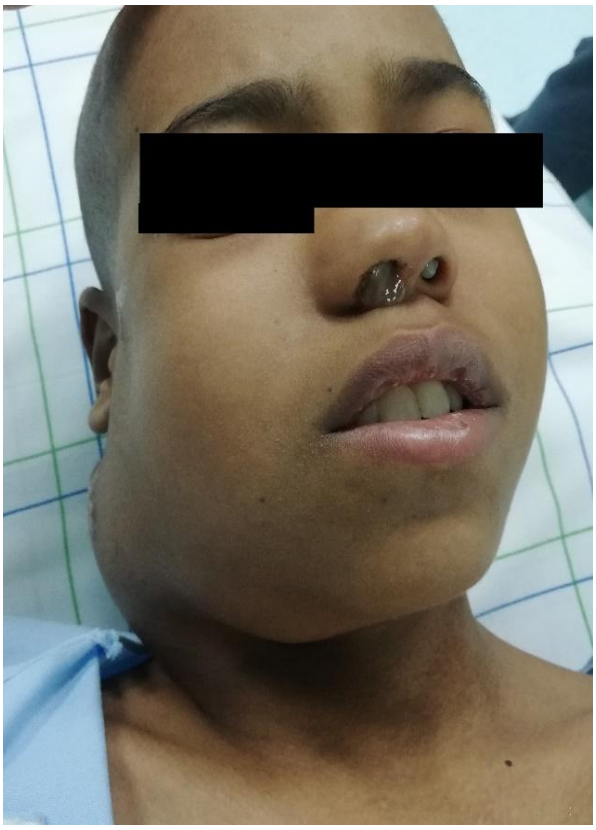

Fig-1: Tumor occupying both nasal cavities with right cervical metastasis

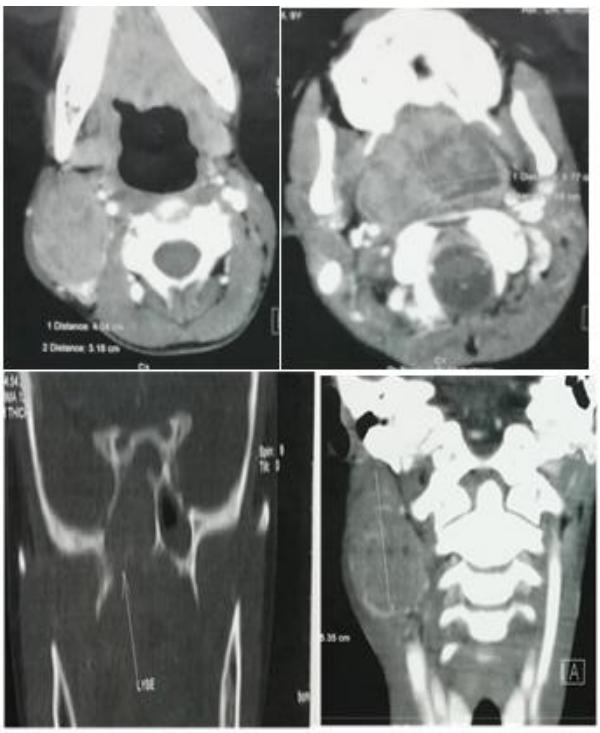

Fig-2: CT images before chemotherapy

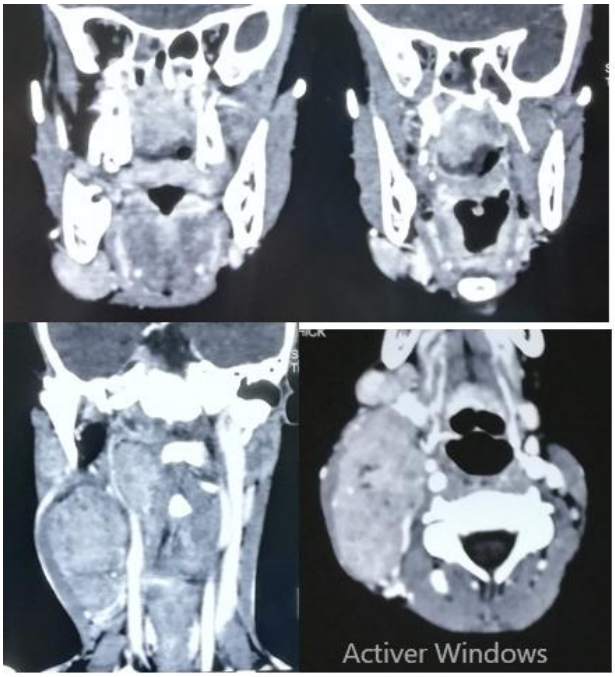

Fig-3: CT images after chemotherapy

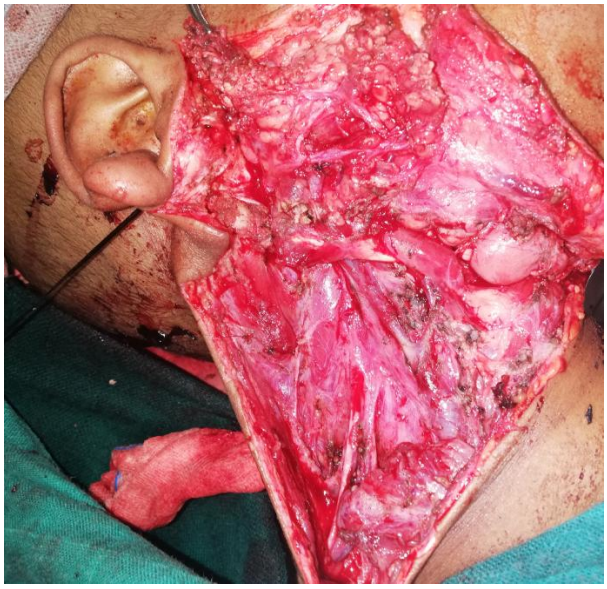

Fig-4: Right parotid and cervical lymph node dissection; operative picture

DISCUSSION

ENB is a rare malignant tumor of the nasal sinus cavities. Its development is thought to originate from olfactory neuroepithelial cells located at the top of the olfactory slits.

These tumors usually present during the sixth and seventh decades, but cases have also been reported in children and in elderly subjects over the age of 80 years [5, 7]. The absence of significance prognostic of these factors on survival, as reported in the literature, is probably due to the very small sample sizes of published series. Similarly, no risk factor for the

pathogenesis of these tumours has ever been identified [8].

Its mode of disclosure is most often nasal obstruction (70%) or unilateral epistaxis (46%) [9]. In some cases, it can manifest as headache, rhinorrhea, anosmia, lacrimation or visual disturbances associated with sinus symptomatology (8-10% of cases) [9]. In our case, the initial symptomatology was the simultaneous appearance of nasal obstruction and a laterocervical mass.

ENB tends to be locally aggressive and metastasizes distally, resulting in decades-delayed recurrences [3, 4, 8, 10-13] with metastatic diseas around 20% of the time but this statistic has been reported as high as 48% [7, 8, 14-20]. The typical sites of metastasis are cervical lymph nodes, lungs, and bones [14, 21, 22]. Less common sites are the liver, mediastinum, adrenal gland, ovary, spleen, parotid glands, central nervous system, and spinal epidural space [14]. Along with metastasis, patient may present with SIADH (syndrome of inappropriate antidiuretic hormone) with dilutional hyponatremia or excess ectopic adrenocorticotropic hormone production leading to Cushing syndrome, although these are rare [23], in our patient there were parotid metastases then cervical lymph nodes united then bilateral, ENB staging can be evaluated according to two classifications: the Kadish classification modified by Morita and the Dulguerov TNM classification [9, 22] .

Computed tomography (CT) and magnetic resonance imaging (MRI) with and without contrast is the first line for evaluation of ENBs [3, 14, 24-27]. Before imaging, it is imperative to obtain a complete medical history, thorough physical examination, and a full record of any other comorbid medical conditions that may alter the treatment plan [14]. The first step is to obtain a full assessment by an otolaryngologist to determine the extent of the disease as well as provide a diagnostic biopsy for histologic grading [14]. A full neurologic and ophthalmologic examination before any extensive surgery can provide a baseline to determine any residual side effects from treatment [14]. Typically, imaging studies show one of the most characteristic findings: a dumbbell-shaped mass extending across the cribriform plate [23, 3, 25]. Where as the waist of the dumbbell is at the cribriform plate, the upper portion is in the anterior cranial fossa and the lower portion is in the nasal cavity [23, 3, 25, 28]. The presence of peritumoral cysts at the tumor-brain interface is another characteristic imaging feature of ENBs [23, 28]. The CT scan is a helpful to identify the lesion but more importantly, it can depict bony erosion of the cribriform plate, orbit, and air sinuses [3, 25, 26]. Typically, on CT, a homogenous mass with necrotic non enhancing areas is observed [3, 25, 26]. The MRI scan helps indicate the extent of soft tissue involvement [3], It is ideal to obtain both scans before any surgical treatment because ENBs can have significant soft tissue involvement that cannot be properly identified on CT scan only. Contrast MRI can help differentiate the tumor from obstructed secretions in the paranasal sinuses, determine meningeal and extradural invasion, and identify the extent of perineural involvement [3, 25, 26].

Histological diagnosis is not difficult if the tumor is well differentiated. It is then composed of small homogeneous and uniform cells with round or oval nuclei, with rosette or pseudo-rosette formations. When the tumor is little or undifferentiated, composed of small hyperchromatic and anaplastic cells, the diagnosis requires immunohistochemical studies with the search for a positive labeling for synaptophysin (> 90%) [29], NSE (neuron specific enolase,> 90%) [29], chro-mogranin, CD56, and lack of expression of muscle markers (desmin, vimentin, actin, myogenin) and markers of epithelial cell differentiation (cyto-keratin) or common leukocytic antigen. The diagnosis sometimes requires the use of electron microscopy which reveals very dense neurosecretory granules in the cytoplasm and nervous processes [29]. Hyams et al., [30] classified ENB into four histopathological stages (I to IV) based on the preservation of lobular architecture, mitotic index, nuclear polymorphism, fibrillar matrix, rosettes and lanecrosis.

There is no agreed standard-of-care treatment algorithm for ENBs. Recent technological advances, improved surgical technique, and better understanding of the disease process has forced physicians to constantly develop new treatment plans, which has made it difficult to assess the effectiveness of each individual treatment [14]. Recently, the interest has shifted from subtotal and total resections to endoscopic transnasal/endonasal procedures with adjuvant radiotherapy or stereotactic radiosurgery. Adjuvant use of radiation and chemotherapy has also shown some benefits. Indeed, surgery remains the first-line treatment for these tumors, and appears in meta-analyzes as an independent factor of progression-free survival and overall survival [32]. The surgical technique depends on the initial tumor stage. For localized tumors, the transfacial route is currently preferred [32]. However, the development of endoscopic surgery over the past decade has enabled comparable carcinological results to be obtained, limiting surgical morbidity and the duration of hospitalization, in particular for stage T1 or T2 tumors [33]. In meta-analyzes, endoscopic resection seems to have better results in terms of overall survival than transfacial surgery. These results should be predicted with caution as studies using transfacial surgery are significantly older than those using endoscopic surgery. This fact may partly explain the more modest results [31].

In the event of a locally advanced tumor (invasion of the orbit or the anterior cerebral fossa), a

mixed surgical approach, by transfacial or endoscopic and neurosurgical route is preferable. Despite the absence of studies of a satisfactory level of evidence, adjuvant radiotherapy is currently considered as standard treatment after surgery, whether histologically complete or not [34].

On the other hand, exclusive radiotherapy should not be used, except in the event of an absolute contraindication to surgery, due to the significantly lower local control and specific survival rate compared to the joint approach [34, 7].

Chemotherapy has shown promising results in patients with high-grade (Hyams grade III or IV) tumors compared with low-grade tumors [14, 36]. In patients with high-grade stage C ENB, adjuvant chemotherapy after complete resection has been beneficial [14, 35]. However, lack of a standardized treatment regimen and small sample sizes in previous studies have made it difficult to provide definitive recommendations for treatment [14]. Theoretically, the similar chemotherapy regimens used for neuroblastomas, small-cell lung carcinoma, and primitive neuroectodermal tumors have been used for ENBs because of their similar histologic and ultrastructural features [12, 21, 36].

CONCLUSION

ENB is a rare malignant tumor originating in the olfactory neuroepithelium that can present with intracranial involvement; strict cases require a multidisciplinary treatment plan. Given its malignant histopathology, metastases are a concern; the most common area for metastasis is the cervical lymph nodes. Thus, we recommend performing head and neck imaging at the very least for appropriate staging. Tumors are chemosensitive and radiosensitive and the literature supports both neoadjuvant and adjuvant therapies with surgery to reduce the tumor burden and provide symptomatic relief from compression of adjacent structures.

Declaration of links of interest

The authors declare that they have no links of interest.

REFERENCES

1. Rimmer J, Lund VJ, Beale T, Wei WI, Howard D. Olfactory neuroblastoma: a 35-year experience and suggested follow-up protocol. Laryngoscope. 2014; 124:1542-1549.
2. Elkon D, Hightower SI, Lim ML, Cantrell RW, Constable WC. Esthesioneuroblastoma. Cancer. 1979;44:1087-1094.
3. Papacharalampous GX, Kotsis GP, Vlastarakos PV, Papadopoulou EP, Saravakos PK, Davilis DI. Olfactory neuroblastoma treated by minimally invasive endoscopic resection and postoperative adjuvant radiotherapy: a representative case and an updated review of the literature. J Cancer Ther. 2012;3:1074-1079.
4. Bradley PJ, Jones NS, Robertson I. Diagnosis and management of esthesioneuroblastoma. Curr Opin Otolaryngol Head Neck Surg. 2003;11:112-118.
5. Broich G, Pagliari A, Ottaviani F. Esthesioneuroblastoma: a general review of the cases published since the discovery of the tumour in 1924. Anticancer Res. 1997;17:2683-2706.
6. Malouf GG, Casiraghi O, Deutsch E, Guigay J, Temam S, Bourhis J. Low- and high-grade esthesioneuroblastomas display a distinct natural history and outcome. Eur J Cancer. 2013;49: 1324-1334.
7. Jethanamest D, Morris LG, Sikora AG, Kutler DI. Esthesioneuroblastoma: a population-based analysis of survival and prognostic factors. Archives of Otolaryngology–Head & Neck Surgery. 2007 Mar 1;133(3):276-80.
8. Dulguerov P, Allal AS, Calcaterra TC. Esthesioneuroblastoma: a meta-analysis and review. Lancet Oncol. 2001;2:683-690
9. Dulguerov P, Calcaterra T. Esthesioneuroblastoma: the UCLAexperience 1970—1990. Laryngoscope. 1992;102:843-9.
10. Palejwala SK, Sharma S, Le CH, Chang E, Erman AB, Lemole GM Jr. Complex skull base reconstructions in kadish d esthesioneuroblastoma: case report. J Neurol Surg Rep. 2017;78: e86-e92.
11. Polin RS, Sheehan JP, Chenelle AG, Munoz E, Lamer J, Phillips CD, Cantrell RW, Laws ER, Newman SA, Levine PA, Jane JA. The role of preoperative adjuvant treatment in the management of esthesioneuroblastoma: the University of Virginia experience. Neurosurgery. 1998 May 1;42(5):1029-37.
12. Loy AH, Reibel JF, Read PW, Thomas CY, Newman SA, Jane JA, Levine PA. Esthesioneuroblastoma: continued follow-up of a single institution's experience. Archives of Otolaryngology–Head & Neck Surgery. 2006 Feb 1;132(2):134-8.
13. Komotar RJ, Starke RM, Raper DM, Anand VK, Schwartz TH. Endoscopic endonasal compared with anterior craniofacial and combined cranionasal resection of esthesioneuroblastomas. World Neurosurg. 2013;80:148-159.
14. Sheehan J, Payne R. Esthesioneuroblastomas. In: Winn HR, ed. Youmans and Winn Neurological Surgery E-Book. Elsevier Health Sciences; 2016, New York, NY:1284-1292.
15. Van Gompel JJ, Giannini C, Olsen KD, Moore E, Piccirilli M, Foote RL, Buckner JC, Link MJ. Long-term outcome of esthesioneuroblastoma: hyams grade predicts patient survival. Journal of neurological surgery. Part B, Skull base. 2012 Oct;73(5):331-336.

16. Cantrell RW, Ghorayeb BY, Fitz-Hugh GS. Esthesioneuroblastoma: diagnosis and treatment. Ann Otol Rhinol Laryngol. 1977;86:760-765.
17. Bailey BJ, Barton S. Olfactory neuroblastoma. Management and prognosis. Arch Otolaryngol. 1975;101:1-5.
18. Kadish S, Goodman M, Wang CC. Olfactory neuroblastoma. A clinical analysis of 17 cases. Cancer. 1976;37:1571-1576.
19. Levine PA, McLean WC, Cantrell RW. Esthesioneuroblastoma: the University of Virginia experience 1960-1985. Laryngoscope. 1986;96:742-746.
20. Levine PA, Gallagher R, Cantrell RW. Esthesioneuroblastoma: reflections of a 21-year experience. Laryngoscope. 1999;109:1539-1543.
21. Richmond CM, Parmley RT. Cytogenetic and molecular evaluation of clinically aggressive esthesioneuroblastoma. Am J Pediatr Hematol Oncol. 1991;13:62-70.
22. Morita A, Ebersold MJ, Olsen KD, Foote RL, Lewis JE, Quast LM. Esthesioneuroblastoma: prognosis and management. Neurosurgery. 1993; 32:706-714 [discussion: 714-715].
23. Gandhoke CS, Dewan A, Gupta D, Syal SK, Jagetia A, Saran RK, Meher R, Srivastava AK, Singh D. A rare case report of mixed olfactory neuroblastoma: carcinoma with review of literature. Surgical neurology international. 2017;8:83.
24. Lund VJ, Stammberger H, Nicolai P, Castelnuovo P, Beal T, Beham A, Bernal-Sprekelsen M, Braun H, Cappabianca P, Carrau R. European position paper on endoscopic management of tumours of the nose, paranasal sinuses and skull base. Rhinol Suppl. 2010 Jun 1;22(2):1-43.
25. Pickuth D, Heywang-Kobrunner SH, Spielmann RP. Computed tomography and magnetic resonance imaging features of olfactory neuroblastoma: an analysis of 22 cases. Clin Otolaryngol Allied Sci. 1999;24:457-461.
26. Yousem DM, Oguz KK, Li C. Imaging of the olfactory system. Semin Ultrasound CT MR. 2001; 22:456-472.
27. Kairemo KJ, Jekunen AP, Kestila MS, Ramsay HA. Imaging of olfactory neuroblastomaean analysis of 17 cases. Auris Nasus Larynx. 1998;25:173-179
28. Tseng J, Michel MA, Loehrl TA. Peripheral cysts: a distinguishing feature of esthesioneuroblastoma with intracranial extension. Ear Nose Throat J. 2009;88:E14.
29. Thompson LD. Olfactory neuroblastoma. Head Neck Pathol. 2009;3:252-9. [Epub 2009 Jul 16. Review].
30. Hyams VJ, Batsakis JG, Michaels L. Olfactory neuroblas-toma. Tumors of the upper respiratory tract and ear, 25.Washington DC: Armed Forces Institute of Pathology; 1988.p. 240-8.
31. Devaiah AK, Andreoli MT. Treatment of esthesioneuroblastoma: a 16-year meta-analysis of 361 patients. Laryngoscope. 2009;119:1412–6.
32. König MS, Osnes T, Meling TR. Treatment of esthesioneuroblastomas. Neuro-chirurgie. 2014;60:151–7.
33. Hanna E, DeMonte F, Ibrahim S, Roberts D, Levine N, Kupferman M. Endoscopic resection of sinonasal cancers with and without craniotomy: oncologic results. Arch Otolaryngol Head Neck Surg. 2009;135:1219–24.
34. Chao KC, Kaplan C, Simpson JR, Haughey B, Spector GJ, Sessions DG, Arquette M. Esthesioneuroblastoma: the impact of treatment modality. Head & Neck: Journal for the Sciences and Specialties of the Head and Neck. 2001 Sep;23(9):749-57.
35. Porter AB, Bernold DM, Giannini C, Foote RL, Link MJ, Olsen KD, Moynihan TJ, Buckner JC. Retrospective review of adjuvant chemotherapy for esthesioneuroblastoma. Journal of neuro-oncology. 2008 Nov;90(2):201-4.
36. Goldsweig HG, Sundaresan N. Chemotherapy of recurrent esthesioneuroblastoma. Case report and review of the literature. Am J Clin Oncol. 1990; 13:139-143.