

Nasal Sinus Esthesioneuroblastoma: A Case Report

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Abstract

Esthesioneuroblastoma (ENB) is a rare malignant neoplasm originating from the olfactory epithelium located in the upper part of the nasal cavities. The treatment is not yet well codified; surgery and external radiotherapy were the standard treatment. The prognosis of these malignant tumors is similar to that of ethmoid adenocarcinomas and better than that of squamous carcinomas of the facial bone structure. 5-year overall and disease-free survival averaged 45%. We reported here a case of a 20-year-old patient treated with neoadjuvant chemotherapy, surgery, and then adjuvant chemotherapy.

Keywords

Esthesioneuroblastoma; Nasal-sinus tumor; Chemotherapy

to the hospital with nasal obstruction, epistaxis, and nasal swelling evolving for 05 months (**Figure 1**). A facial Magnetic Resonance Imaging showed a tumor of the left nasal cavity and ethmoidal cells measuring 47 x 71 x 89 mm. This tumor process fills the nasal cavities, ethmoidal cells, frontal sinuses, and the anterior level of the base of the skull. The biopsy with immunohistochemistry (the Anti-bodies: anti-GFAP negative, anti-Chromogranin A positive, anti-Synaptophysin positive, anti-CD99 negative, anti-Cytokeratin positive and anti-Protein S-100 positive) is in favor of an esthesioneuroblastoma. The tumor was classified stage C of Kadish.

He received 03 cycles of neo-adjuvant chemotherapy based on cisplatin Etoposide (every 03 weeks) followed by surgical excision (R2 excision). Three cycles of the same chemotherapy regimen was administered postoperatively. The inter-course intervals in chemotherapy were respected. The patient was referred to our health facility for external radiotherapy (Intensity Modulated Radiotherapy-IMRT) and the tumor progressed before his arrival. The tumor invades the brain parenchyma and he was recused for the curative radiotherapy (**Figure 2**). He is then referred for palliative chemotherapy.

Introduction

Esthesioneuroblastoma (ENB) is a rare malignant neoplasm originating from the olfactory epithelium located in the upper part of the nasal cavities [1]. Molecular studies indicate that the basal progenitor cells of the olfactory epithelium are the cause of esthesioneuroblastoma. The histopathological diagnosis remains difficult and relies on the results of the expression of antigens detected through a panel of antibodies by immunohistochemistry [1]. The treatment is not yet well codified; external surgery and radiotherapy were the standard treatment. The use in recent years of endoscopic endo-nasal surgery and chemotherapy appears promising [2]. We report here a case of ENB treated with chemotherapy and surgery and which progressed during treatment.

Observation

A 20-year-old young boy with no prior medical history, admitted

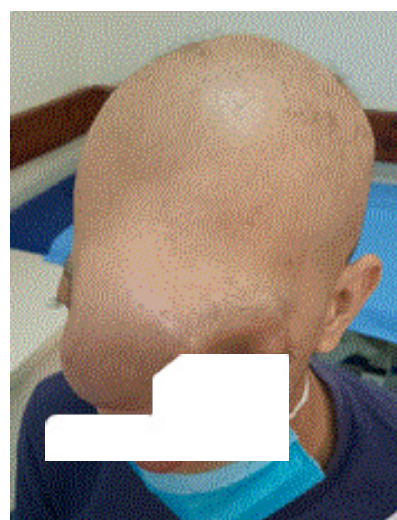


Figure 1: Image of the mass.

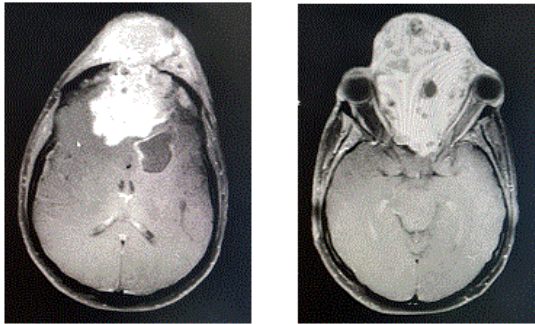


Figure 2: Cerebral MRI, showing intracranial invasion of the tumor.

Discussion

Esthesioneuroblastoma was described for the first time by Berger and Luc in 1924 [3]. Less than 1000 cases in the world have been published during the last twenty years, with less than 1 case per million [4]. It is a rare and clinically variable malignant tumor of the nasal sinus and the base of the skull [5,6]. Symptoms of the tumor are related to its location and generally include: epistaxis, nasal obstruction, olfactory, and ophthalmic disorders as well as craniofacial pain [1]. From a clinical point of view, this tumor generally grows slowly and mainly locally, but it is important to be aware of the possibility of rapid progression with diffuse bone metastases [7]. The first historical classification proposed by Kadish in 1974 is based on clinical extension [8], this classification was then modified by Foote [9]. Then, Dulguerov proposed in 1992 a new classification closer to the TNM classifications usually used [10].

As with all other malignant tumors, the therapeutic arsenal is based on surgery, radiotherapy, and chemotherapy. The standard treatment is anterior craniofacial resection with postoperative irradiation. The role of chemotherapy is discussed, but is generally for the most advanced cases and used in the neo-adjuvant and/or postoperative setting with irradiation [6,7,11,12]. However, recent literature supports that endoscopic resection correlates with similar oncologic control rates, compared to open surgery when basic oncologic surgical principles are maintained [13].

For radiotherapy, a dose of 55.5 Gy is recommended after complete resection of the esthesioneuroblastoma [9]. Intensity-modulated radiation therapy and endoscopic surgery have reduced morbidity, but the results with these techniques need to be fully evaluated [6]. The prognosis of these malignant tumors is similar to that of ethmoid adenocarcinomas and better than that of squamous carcinomas of the facial bone structure [7]. 5-year overall and disease-free survival averaged 45% [10].

We have reported a case of a patient treated with neo-adjuvant chemotherapy, surgery, and then adjuvant chemotherapy who progressed under treatment and therefore did not receive curative radiotherapy.

Conclusion

ENB is a rare rhino-sinus tumor that has the particularity of being able to recur very late. The first-line treatment for resectable tumors is complete resection surgery followed by radiotherapy. The place

of chemotherapy has yet to be determined. While it now seems to be proven that neo-adjuvant chemotherapy does not provide any benefit in terms of survival, adjuvant chemotherapy, especially for stage C or D tumors, could represent the next therapeutic advance in ENB.

Ethics approval and consent to participate

Written informed consent was obtained from the patient and his family for publication of this case reports any accompanying images.

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