

Article

Massive Esthesioneuroblastoma of middle turbinate

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Abstract. Esthesioneuroblastoma involving the anterior skull base is a rare malignant tumor derived from the olfactory epithelium, about its treatment there is currently no universally accepted protocol. Aim of this paper is to illustrate the exceptional finding of a ENB in the anterior horn of right middle turbinate.

Keywords: Esthesioneuroblastoma; olfactory cells; nasal fossa.

Introduction

For the first time described in 1929 by Luc and Berger, Esthesioneuroblastoma (ENB) is a rare distinctive malignant tumor of the sinonasal region, deriving from neuroendocrine cells of olfactory epithelium (1,2,3,4,5,6,7). ENB is well known for its rarity, in fact about 1000 cases are described in literature since its first detection (2,3,7). ENB presents a large variety of symptoms and rapid progression. Frequently detection of ENB is not immediate, this delay of diagnosis makes difficult to reach a consensus on an ideal staging system, prognostic evaluation and best treatment modalities (1,4).

The prognosis of ENB is generally determined by a combination of clinical and pathologic factors. Factors with a negative impact on prognosis include advanced stage, younger age, high grade of differentiation, positivity of resection margins, regional nodal and distant metastases, high proliferation indices, p53 overexpression (2,3,8).

The Hyams grading system has been extensively used to predict prognosis in patients with ENB. However, most studies showing prognostic correlation group grading into I/II versus III/IV, essentially comparing low versus high grade. In addition, these studies include patients with variable treatment regimens, including some that were treated with chemoradiation alone (9,10). Aim of this paper is to illustrate the exceptional finding of a ENB in the anterior horn of right middle turbinate.

Case

In April 2013 a caucasian female aged 46 was referred to our observation at Maxillo Facial surgery department of S. Andrea Hospital, Second Faculty of Medicine and Surgery of University of Rome "la Sapienza". Patient was complaining about a reduction of airflow during nasal respiration. A reduction of right noethrill was clearly visible with anterior rhinoscopy.

The enhanced CT scan showed a lesion of the anterior horn of the right middle turbinate, which extended to the nasopharynx. Patient underwent to surgical treatment on late april, the formation has been removed with the entire middle turbinate and the histopathological exam revealed how the solid mass were a ENB completely excised. Patient was dismissed in fifth postoperative day with no complications.

Discussion and conclusion

ENB involving the anterior skull base is a rare malignant tumor derived from the olfactory epithelium, about its treatment there is currently no universally accepted protocol (13). The gold standard of surgical treatment is currently craniofacial resection, which allows efficient removal of the tumor but entails significant morbidity. To reduce morbidity combined with good visualization of tumor limits removal, endonasal endoscopy resection (EER) has developed (11).

Combination of surgery and radiotherapy (either conventional radiotherapy or stereotactic radiosurgery), with or without chemotherapy is considered to be the standard of care for primary site disease by the majority of researchers. (12)

Rimmer in his 35-years retrospective study, observes that olfactory neuroblastoma most commonly recures within the first 4 years (13); in his experience he observed just one case of relapse after 19 years after surgical procedure.

Even according with Rimmer, follow-up regime consists lifelong follow-up with both clinical examination and serial imaging, including the neck and entire intracranial compartment.

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