Auris Nasus Larynx xxx (2018) xxx–xxx

Contents lists available at ScienceDirect

Auris Nasus Larynx

journal homepage: www.elsevier.com/locate/anl

Case report

Treatment of pediatric esthesioneuroblastoma with smell preservation

Giandomenico Maggiore, Maria Silvia Lazio *, Oreste Gallo

Clinic of Otolaryngology, Head and Neck Surgery, Department of Translational Surgery and Medicine, University of Florence, Italy

ARTICLE INFO

Article history:
Received 31 October 2017
Accepted 6 February 2018
Available online xxx

Keywords:
Paranasal tumors
Olfactory neuroblastoma
Esthesioneuroblastoma
Smell preservation
Sinonasal malignances

ABSTRACT

Olfactory neuroblastoma is a rare malignant tumor of neuroectodermal origin and represents the most common cancer of the nasal cavity in pediatric age. The gold standard of treatment consists of en bloc resection, numerous studies have shown as the endoscopic approaches permit good control of the disease improving the quality of life after the treatment. Herein we describe the case of a 13-year-old patient referred to our outpatient clinic with a polypoid multi-lobed lesion occupying the left nasal cavity and imaging that confirmed a left-sided nasal mass without cribriform plate involvement (Kadish B). We performed an unilateral endoscopic resection with transnasal craniectomy and anterior skull base reconstruction with a flap from the contralateral nasal septum based on the septal branches of the anterior and posterior ethmoidal arteries (Septal Flip Flap, SFF), that provided a faster healing process with reduction of nasal crusting, improvement in the quality of life of patient in the postoperative period and the preservation of the contralateral olfactory bulb that has allowed to save the smell. This treatment strategy of pediatric esthesioneuroblastoma was analyzed in the context of the current literature.

1. Introduction

Olfactory neuroblastoma (ON), also known as esthesioneuroblastoma (ENB) was described by Berger in 1924, who described an "esthesioneuroepitheliome olfactif" [1]. Since the first description about 1000 cases have been reported in the world literature [2]. It represents about 3%–5% of all malignant nasal tumors [3] and has a bimodal age distribution, between 10 and 20 years and between 50 and 60 years. In pediatric age, the estimated incidence of ON is 0.1/100.000 children up to 15 years of age, but it is the most common cancer of the nasal cavity [4,5]. Tumor etiopathogenesis of ON is not yet clear, there are no known etiologic agent(s) for human olfactory neuroblastoma demonstrated. The main presenting symptoms are unilateral nasal obstruction (70%) and epistaxis (46%); less common manifestations include anosmia, headache, pain, excessive lacrimation and ocular disturbances, these symptoms are related to the site and local invasion of the tumor. Diagnosis is often made late because they are asymptomatic or produce nonspecific symptoms in their early stages, the mean delay between the appearance of the first symptom and the diagnosis being 6 months [6]. ON does not have any specific radiologic appearance and when seen, it is a homogenous soft-tissue mass in the nasal vault, with uniform and moderate contrast enhancement. Computed tomography (CT) images are essential for correct staging and they should be carefully examined for erosion of the lamina papyracea, cribriform plate, and fovea ethmoidalis. Magnetic resonance imaging (MRI) enables a better estimate of tumor spread into surrounding soft-tissue areas and can differentiate mucus from tumor, also showing any possible intraorbital or intracerebral extension. Regional and metastatic dissemination occurs, with 8–20% of patients having

* Corresponding author at: Clinic of Otolaryngology, Head and Neck Surgery, University of Florence, via Largo Brambilla, 3-50134 Firenze, Italy.
E-mail address: silvia.la89@gmail.com (M.S. Lazio).

https://doi.org/10.1016/j.anl.2018.02.003
0385-8146/

clinical or occult cervical nodal involvement at presentation [7]. Kadish et al. [8] were the first to propose a staging classification, of 3 stages (A–C) for ON, which was later modified by Morita [9], who added stage D indicating metastatic disease. Optimal treatment is still discussed controversially. In adults, Kadish Stage and Hyam's histological grade have been shown to predict prognosis [10–12]. This has not been validated in children that generally present with more advanced stage disease than in adults, however, their prognosis is better. Most children are treated based on reviews of adult cases and series and based on the findings of a limited number of studies, the gold standard for treatment of ON is craniofacial resection (CFR) followed by adjuvant radiotherapy and/or chemotherapy [13]. Most recently, with the advent of endoscopic technologies, use of a transnasal surgical approach has achieved comparable outcomes to traditional open craniofacial resection [14–16]. Although combined surgery and radiation therapy has produced the best outcomes in adults, in children it can lead to significant long-term morbidity. Herein we describe the pediatric case of a patient with ON treated by radical endoscopic surgery alone with preservation of the contralateral olfactory bulb in order to preserve olfaction.

2. Case report

A 13-year-old male referred to our outpatient clinic with a 3 months history of recurrent epistaxis from the right nostril, unilateral nasal obstruction and congestion symptoms without pain or ocular disturbances. Parents referred a normal psychophysical growth, no prior cranio-facial trauma, surgery or hospitalizations and a non-significant family medical history. Antibiotics, systemic anti-inflammatory and topical therapy were before prescribed without benefit. The University of Pennsylvania Smell Identification Test (UPSIT) [17] was administered to assess olfactory function that resulted normal with a score of 35. The video-endoscopic examination with 0° rigid telescope showed a polypoid multi-lobed lesion occupying the right nasal cavity, of dark red color and easily bloody with mechanical stimulation. The base of implant of the lesion was impossible to identify. It was required a MRI plus gadolinium that revealed a lesion with marked enhancement occupying the right nasal cavity and involved the medial wall of the maxillary sinus, extending backwards to the posterior ethmoid and the frontal recess. There was no evidence of orbital, meningeal and cerebral invasion (Fig. 1). CT scan confirmed the presence of the mass occupying the right nasal cavity with ipsilateral involvement of maxillary and ethmoid sinuses, the mass was in contact with the medial orbital wall without evidence of cribriform plate erosion. Because the mass showed a marked enhancement both on CT and MRI, the patient underwent a video endoscopic assisted biopsy under general anesthesia in order to avoid an important bleeding in outpatient setting, in the suspect of a vascular mass. Intraoperative biopsy showed malignant features of the lesion. After fixation, histopathological and immunohistochemical examination were diagnostic for ON with Hyams grade II differentiation. Positron Emission Tomography (PET-CT) scan was performed, with evidence of high metabolisms areas in the right nasal cavity and in a single right retro-angolo- mandibular lymph node. This latter was examined with a neck ultrasonography that showed a reactive morphologic features of the lymph node. The ON was stage B, according to Kadish classification and T2 according to Dulguerov TNM staging [18]. Treatment management was discussed in our Pediatric Tumor Board with indication to surgical endoscopic resection. The patient underwent an endoscopic sinuses surgery, as first step we realized a debulking of the sinonasal tumour in order to assess the extent of disease and its origin and exclude any involvement of the nasal septum. Then the base of implant of the tumor was removed using a subperiosteal dissection plane, with the centripetal technique obtaining a clean macroscopic surgical field. Intraoperative frozen sections of the contralateral nasal septum mucosa were negative for malignancy. Once the bony of the anterior skull base was exposed, the skull base was drilled out using a diamond burr and last, after resection of the ethmoidal roof and the fovea ethmoidalis, we removed the dura mater of the olfactory cleft and the olfactory bulb (Fig. 2a). The dura was then circumferentially incised with angled scissors and resected at a safe distance from the suspected area of tumour invasion. We verified the surgical field with 11 frozen sections to minimize the risk of recurrence, resulting all free of disease. In our patient, it was possible to anatomically spare the contralateral olfactory cleft. The resulting skull base defect was reconstructed transnasal in a multilayer fashion using free grafts of iliotibial tract. in detail, the first layer was placed in the intradural gap and the second in the epidural one. The duraplasty was then resurfaced using the septal flip flap (SFF), a pedicled flap based on the septal branches of the contralateral ethmoidal arteries. After the preparation of the flap according to the technique described by Battaglia et al. [19] was made a superiorly hinged and freely rotated to cover the anterior skull base defect as a third layer (Fig. 2b). The flap is then properly fixed with fibrin glue and absorbable hemostat and the nasal cavities are packed bilaterally with polyvinyl alcohol sponge for about 48 h. The operation was free of complications, the subsequent pathology report confirmed that all margins were free from disease, in particular we obtained 22 margins to valuate for the definitive examination. Immediate Postoperative MRI and a CT scan revealed no evidence of residual tumor and the preservation of the left olfactory bulb (Fig. 3), and after a new discussion in our Pediatric Tumor Board no adjuvant radiation therapy was prescribed. During clinical follow up olfactory function was investigated by an interview (preserved or loss) and during the last clinical control a new UPSIT test was assessed and shows a normal smell function with a score of 38 at 24 months after surgical treatment. The patient remained disease free at last follow-up of 24 months.

3. Discussion

Pediatric ON is a rare disease that usually affects the adolescent age group and due to its rarity, there are no randomized, controlled studies investigating optimal treatment guidelines. First-line treatment for ON is CFR with postoperative radiation therapy, combining a bifrontal craniotomy and transfacial approach to achieve true en bloc resection performed

Please cite this article in press as: Maggiore G, et al. Treatment of pediatric esthesioneuroblastoma with smell preservation. Auris Nasus Larynx (2018), https://doi.org/10.1016/j.anl.2018.02.003