International Journal of Pediatric Otorhinolaryngology 87 (2016) 44–49

Contents lists available at ScienceDirect

International Journal of Pediatric Otorhinolaryngology

journal homepage: http://www.ijporlonline.com/

Review article

Pediatric sialoblastoma: Evaluation and management

Alexandria L. Irace [a], Eelam A. Adil [a, b], Natasha M. Archer [c], Victoria M. Silvera [d], Antonio Perez-Atayde [e], Reza Rahbar [a, b, *]

[a] Department of Otolaryngology and Communication Enhancement, Boston Children's Hospital, 300 Longwood Avenue, Boston, MA 02115, USA
[b] Otolaryngology, Harvard School of Medicine, 25 Shattuck Street, Boston, MA 02115, USA
[c] Pediatric Hematology/Oncology, Dana-Farber/Boston Children's Cancer and Blood Disorders Center, 450 Brookline Avenue, Boston, MA 02215, USA
[d] Department of Radiology, Boston Children's Hospital, 300 Longwood Avenue, Boston, MA 02115, USA
[e] Department of Pathology, Boston Children's Hospital, 300 Longwood Avenue, Boston, MA 02115, USA

ARTICLE INFO

Article history:
Received 9 March 2016
Received in revised form
27 April 2016
Accepted 28 April 2016
Available online 29 April 2016

Keywords:
Pediatric sialoblastoma
Salivary gland tumor
Parotid neoplasm
Salivary embryoma
Congenital basal cell adenoma
Congenital hybrid basal cell adenoma-adenoid cystic carcinoma

ABSTRACT

Objectives: Sialoblastoma is a rare congenital salivary gland tumor of epithelial origin. The objectives of this study are to review the literature regarding clinical presentation of sialoblastoma, evaluate the effectiveness of various treatment methods, and present guidelines for evaluation and management in the pediatric population.
Data sources:: Case presentation and literature review.
Review methods: A comprehensive search was conducted to identify cases of pediatric sialoblastoma in the English-language literature. The presentation, evaluation, and management of reported cases were analyzed. We also report an invasive and recurrent case in a pediatric patient to highlight the aggressive nature of these lesions.
Results: Sixty-two cases of pediatric sialoblastoma were reviewed. The age at initial presentation ranged from before birth to 15 years. The parotid gland was the most common location (n = 47). Surgical excision was the primary treatment in all patients. Nine patients developed metastatic disease of the lung, lymph nodes, or bone. Almost a third of patients had recurrence and over two thirds of patients were tumor-free for at least 1 year following their last treatment intervention.
Conclusion: Prompt and complete surgical excision should be recommended to prevent local and systemic recurrence of pediatric sialoblastoma. Chemotherapy has also shown promise in several cases, and clinical genomics may shed light on more therapy options. Patients should be closely followed for at least 12 months following diagnosis, or longer depending on the histopathological staging of the tumor.

Contents

Presented at the American Society of Pediatric Otolaryngology, Combined Otolaryngology Spring Meetings, Chicago, IL, May 20, 2016.
* Corresponding author at: Otolaryngology, Harvard School of Medicine, 25 Shattuck Street, Boston, MA 02115, USA. Tel.: +617 355 6462; fax: +617 730 0320.
E-mail address: Reza.rahbar@childrens.harvard.edu (R. Rahbar).

http://dx.doi.org/10.1016/j.ijporl.2016.04.037
0165-5876/

1. Introduction

Sialoblastoma is a rare tumor that arises from the primitive duct epithelium of salivary glands during the perinatal period. Lesions are typically detected by antenatal sonography or by caregivers within the first few weeks of life. It is estimated that 2% to 5% of all salivary gland tumors occur in children, with an annual incidence of 0.8 per million, making sialoblastoma exceptionally uncommon [1,2]. This neoplasm has been reported to be three times as likely to occur in the parotid gland than the submandibular gland [3]. Sialoblastoma is known to be locally infiltrative and may metastasize to the lymph nodes or lungs if left untreated [4–6].

Due to the rarity of sialoblastoma, there is little consensus on its management. In this article, we present a patient with a rapidly growing malignant and recurrent sialoblastoma of the parotid gland. The patient underwent multiple surgical resections and cycles of chemotherapy. We also review the literature to summarize patient presentation, management, and outcomes. Our goal is to propose guidelines for evaluation and management of this rare tumor based on our experience and the prior literature.

2. Methods

A case of a pediatric patient who presented with a large sialoblastoma of the parotid gland is described. A literature search was performed using the online search databases PubMed, EMBASE, CINAHL with Full Text, and Cochrane Library on August 1, 2015. Medical subject headings (MeSH) terms included sialoblastoma, salivary embryoma, congenital basal cell adenoma, congenital hybrid basal cell adenoma, and congenital hybrid basal cell adenoma-adenoid cystic carcinoma. The term “low-grade basaloid adenocarcinoma,” a previously used name for sialoblastoma, was excluded due to overlapping nomenclature with other salivary gland tumors, as defined by the World Health Organization [3]. Search results were restricted to those written in the English language describing pediatric patients (under the age of 21 years of age) and were not limited by year of publication. Separate articles describing the same patient were considered one result.

3. Results

3.1. Case presentation

A 5-month-old male was referred to our institution with a left-sided firm facial mass. The patient was noted at birth to have a 1 cm × 1 cm mass overlying the mandible. This was initially stable and then rapidly grew to 3 cm × 3 cm by 2 months of age. Ultrasound of the mass at this point showed a mildly heterogeneous, well-defined predominantly hypoechoic mass within the left parotid gland with scattered internal hyperechoic foci (Fig. 1) and mild increased vascularity. The mass was initially believed to be an infantile hemangioendothelioma and was monitored by a local otolaryngologist in the patient’s home country. Its continued rapid growth prompted referral to our institution for further management.

The mass on presentation was friable and warm to the touch.

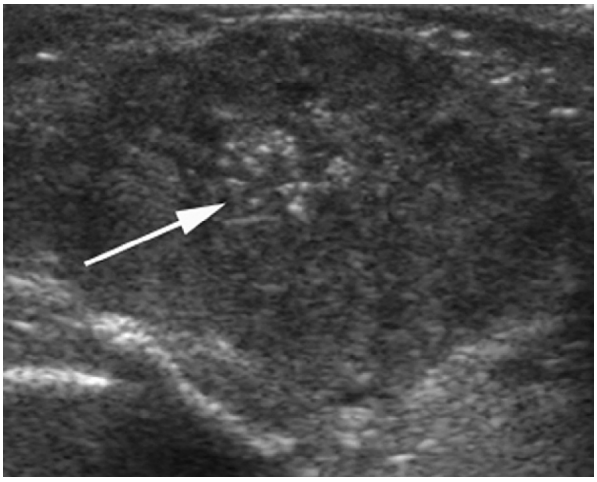

Fig. 1. Ultrasound image shows a circumscribed, hypoechoic mass with scattered hyperechoic foci (arrow).

The patient had dysphagia and left-sided facial nerve paralysis. Magnetic resonance imaging (MRI) revealed a large mass extending intracranially via a widened left foramen ovale into the cavernous sinus. There was also extension into the left external auditory canal/left middle ear cavity, lower eyelid and possibly the superolateral orbit. The mass measured 12.1 cm anterior-posterior by 13.5 cm craniocaudal by 9.5 cm transverse in maximal dimension. Tumor was isointense to muscle on T1-weighted images (T1WIs) (Fig. 2A). T2-weighted images (T2WIs) showed areas of intermediate signal intensity, suggesting components of tumor with a high cytoplasm to nucleus ratio (Fig. 2B) that also corresponded to regions of reduced diffusivity on the ADC map (Fig. 2C), and areas of increased signal consistent with high water content or necrosis. Post-contrast T1WIs showed intense, heterogeneous tumor enhancement (Fig. 3). Contrast-enhanced computed tomography (CT) showed heterogeneous tumor enhancement and bony destruction of the left temporal, sphenoid, pterygoid, maxillary, mandibular, and zygomatic bones, and bony remodeling/thinning of the squamosal portion of the temporal bone (Fig. 4). No areas of bone destruction or flow voids were appreciated on MRI or CT. Chest CT revealed no metastatic disease. Biopsy of the mass was guided by interventional radiology, and pathological examination revealed a malignant tumor consistent with sialoblastoma.

The patient received 5 cycles of neoadjuvant chemotherapy as per the intermediate-risk neuroblastoma regimen (carboplatin, cyclophosphamide/etoposide, and doxorubicin) in an effort to shrink the tumor to a more easily resectable size. The mass shrank to approximately 3 cm × 4 cm following chemotherapy. He underwent aggressive surgical debulking of the extracranial component of the tumor and total parotidectomy. Pathological examination of the resected specimen confirmed the diagnosis of sialoblastoma with multifocal tumor necrosis and infiltration but no evidence of lymphovascular invasion (Fig. 5).

Approximately one month following surgery, disease progression shown by MRI prompted a 6-month course of adjuvant