International Journal of Pediatric Otorhinolaryngology 74 (2010) 942–945

Contents lists available at ScienceDirect

International Journal of Pediatric Otorhinolaryngology

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

Case report

Sialoblastoma of salivary glands in children: Chemotherapy should be discussed as an alternative to mutilating surgery

M. Prigent [a,*], N. Teissier [a], M. Peuchmaur [b], M. El Maleh-Berges [c], P. Philippe-Chomette [d], P. Cardin [e], D. Orbach [f]

[a] E.N.T. Department, Robert Debré-Assistance Publique Hospital, 48 rue Serurier, 75019 Paris, France
[b] Pathology Department, Robert Debré-Assistance Publique Hospital, 48 rue Serurier, 75019 Paris, France
[c] Diagnostic Imaging Department, Robert Debré-Assistance Publique Hospital, 48 rue Serurier, 75019 Paris, France
[d] Surgery Department, Robert Debré-Assistance Publique Hospital, 48 rue Serurier, 75019 Paris, France
[e] Pierre Cadin, Paediatric Bullion Hospital, 78 830 Bullion, France
[f] Paediatric Department, Institut Curie, 26 rue d'Ulm 75005 Paris, France

ARTICLE INFO

Article history:
Received 3 December 2009
Received in revised form 27 January 2010
Accepted 31 January 2010

Keywords:
Sialoblastoma
Salivary gland tumor
Chemotherapy
Children

ABSTRACT

Sialoblastoma is a very rare congenital salivary gland tumor. No consensus has been reached concerning the treatment of this tumor due to its rarity. The treatment of reference is surgery, which can be mutilating, in the case of a locally invasive tumor. The treatment of metastatic disease is also controversial. The authors report a new case of a 6-year-old girl with a progressively growing left parotid mass since birth. The first cytological diagnosis was that of pleomorphic adenoma. Due to local progression, superficial parotidectomy was performed at the age of 3.5 years and revealed a diagnosis of sialoblastoma. Six months later, local recurrence and lung metastasis were treated by neoadjuvant chemotherapy with a very good partial response on the local recurrence and the lung metastasis, allowing complete parotidectomy with sacrifice of the facial nerve. Bilateral lung biopsies after adjuvant chemotherapy showed total necrosis. No recurrence was observed with a follow-up of 1 year.

This case and a review of the literature confirm the very good chemosensitivity of this tumor and argue in favor of neoadjuvant chemotherapy for locally invasive tumors rather than extensive mutilating surgery.

1. Introduction

Sialoblastoma is a very rare congenital salivary gland tumor, only 26 cases are reported [1–19]. This tumor may exist before birth [7,11,13,18], or may be discovered after delivery [6,8,19]. It mainly arises in the parotid gland (16 of the 21 published cases) [3,6,8–11,14–16,18–20], but may also involve the submandibular gland (6 cases) [6,11,12] or even ectopic salivary gland tissue [2]. Due to the rarity of this tumor, no consensus has been reached concerning the treatment of sialoblastoma. The treatment of reference is surgery with complete tumor resection. Local and regional recurrences are frequent despite surgery and surgery can also be mutilating in the case of a locally invasive tumor. The treatment of metastatic disease is also controversial. Some cases of successful chemotherapy have been reported [3,6,9,13,15,21]. Due to the young age of these patients, external beam radiotherapy does not appear to be an appropriate adjuvant treatment option [19]. We present a new case of a 6-year-old girl with left parotid sialoblastoma and lung metastasis successfully treated by neoadjuvant chemotherapy, local conservative surgery and adjuvant chemotherapy.

2. Case report

Born in Algeria, Z. was the first of two children. She was born at term by vaginal delivery. Antenatal ultrasound did not detect any abnormalities. Soon after birth, her parents discovered a small swelling of the left parotid gland. The mass was initially nodular, firm and non-inflammatory. Due to its progressive increase in size, displacing the ear lobe posteriorly, fine needle aspiration was performed, providing results compatible with pleomorphic adenoma. The tumor spontaneously decreased in size and was no longer clinically visible. Consequently, no further examinations were performed.

* Corresponding author. Tel.: +33 612225095.
E-mail address: marinepri@hotmail.com (M. Prigent).

0165-5876/$ – see front matter. Crown Copyright © 2010 Published by Elsevier Ireland Ltd. All rights reserved.
doi:10.1016/j.ijporl.2010.01.026

Four years later, the child presented with a large, firm, heterogeneous, infiltrative and painless mass of the left cheek mass, measuring 3 cm in diameter. The mass caused neck stiffness and inflammation of the skin of the cheek. There were no signs of facial palsy. CT scan showed heterogeneous swelling of the parotid gland with no extension to the pterygoid fossa. No lymph node invasion was observed. Chest X-ray was normal. Superficial parotidectomy was then performed. Histological examination concluded on the diagnosis of sialoblastoma. Surgical resection was considered to be complete.

One year later, a mass recurred in the retro-auricular zone with progressive left facial palsy. On CT scan, the tumor involved the deep parotid lobe.

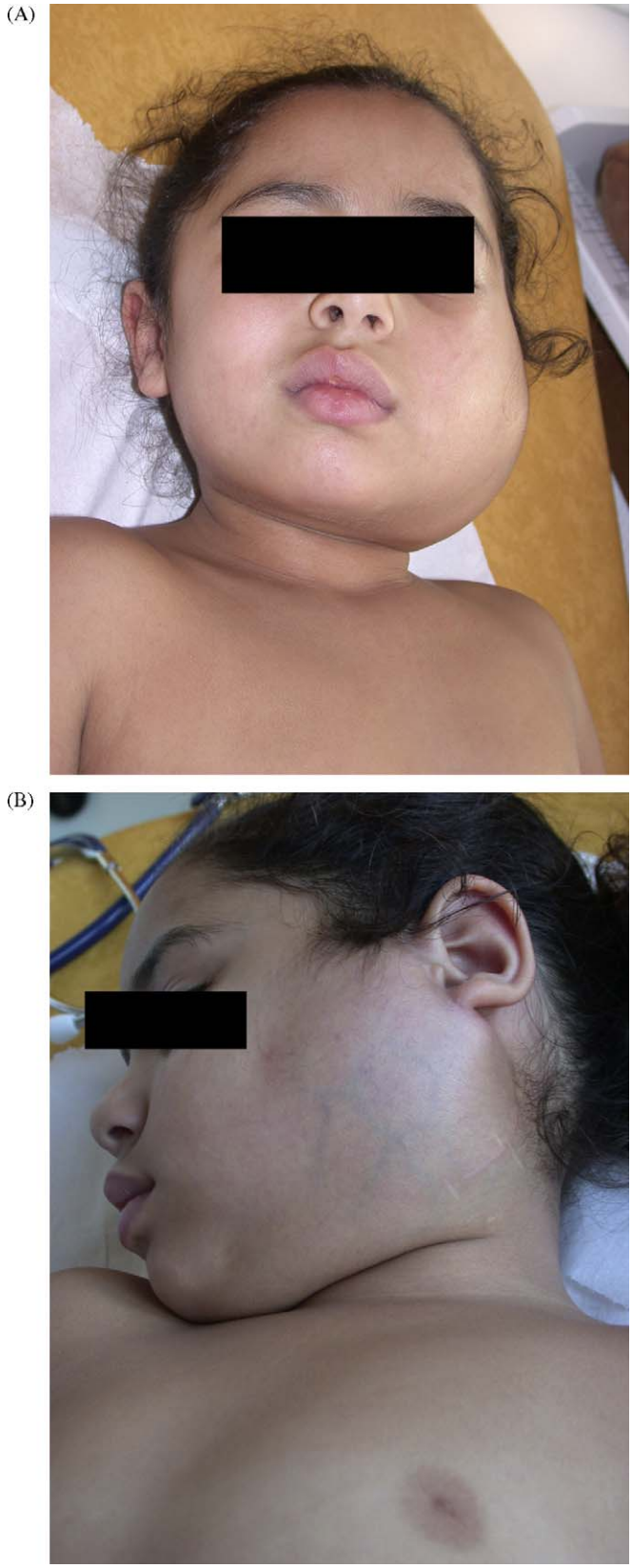

Fig. 1. (A and B) Clinical appearance of local relapse (anterior and lateral view): impressive solid parotid mass with left facial palsy.

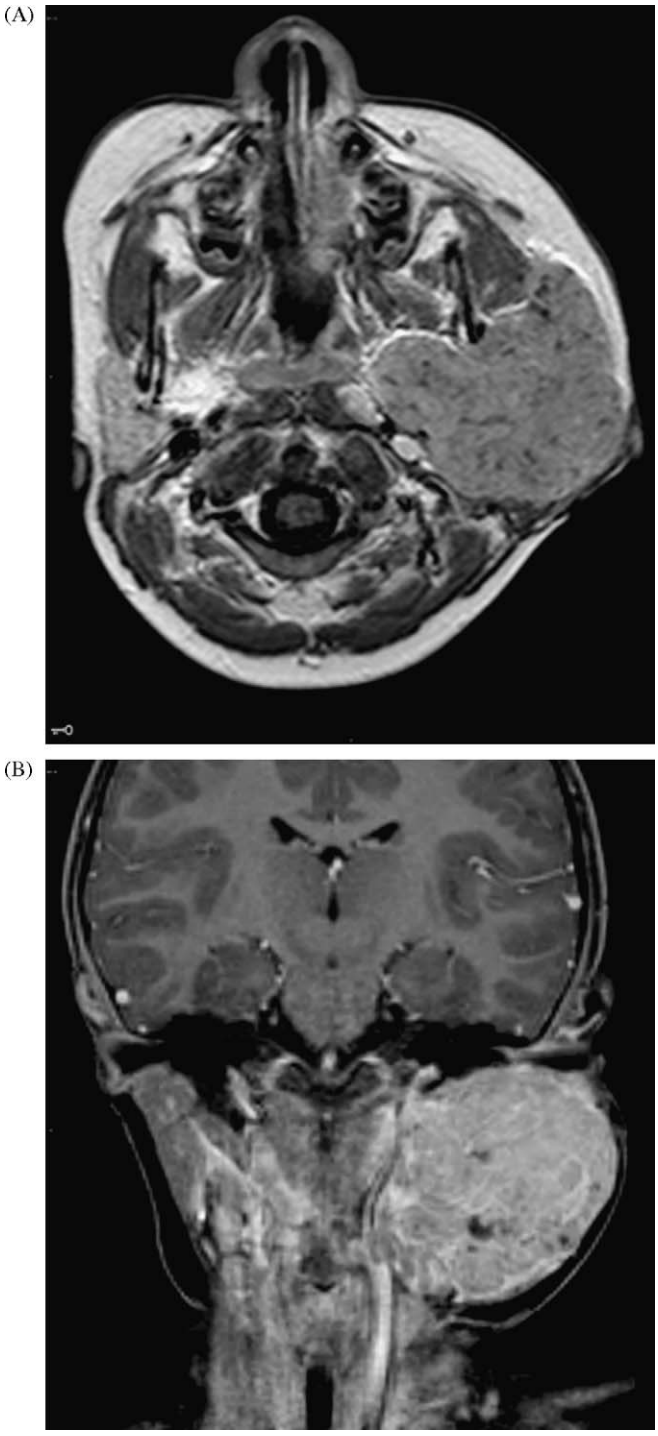

Fig. 2. Magnetic resonance imaging of the head. (A) Contrast-enhanced T1-weighted axial view; well-delineated left parotid swelling with heterogeneous contrast enhancement. (B) Contrast-enhanced T1-weighted frontal view with fat suppression; the left internal carotid is displaced medially by the lesion.