International Journal of Pediatric Otorhinolaryngology 78 (2014) 142–147

Contents lists available at ScienceDirect

International Journal of Pediatric Otorhinolaryngology

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

Case Report, Case Reports

Pediatric laryngeal neurofibroma: Case report and review of the literature

Steven B. Chinn [a], Ryan M. Collar [a,1], John B. McHugh [b], Norman D. Hogikyan [a], Marc C. Thorne [a,*]

[a] Department of Otolaryngology – Head and Neck Surgery, United States
[b] Department of Pathology, University of Michigan Medical School, Ann Arbor, MI, United States

ARTICLE INFO

Article history:
Received 16 July 2013
Received in revised form 29 October 2013
Accepted 30 October 2013
Available online 13 November 2013

Keywords:
Laryngeal neurofibroma
Neurofibromatosis
Pediatric
Transoral CO_2 laser resection

ABSTRACT

Presentation of a case of pediatric laryngeal neurofibroma (LNF) and review of the world literature. Comprehensive review of the world literature using Pubmed and Google scholar. Pediatric LNF was identified in 62 cases reported in the world literature. The most common presenting symptom is stridor and the most common location of the tumor in the larynx is the aryepiglottic fold. Recent reports demonstrate increased utilization of endoscopic resection with reduced need for tracheostomy. Pediatric LNF is a rare disorder. Review of the world literature since 1940 suggests a recent trend away from aggressive open resection and toward more conservative endoscopic resection with excellent functional results.

1. Introduction

Laryngeal neurofibromas (LNF) represent an extremely rare cause of upper airway obstruction in the pediatric patient population. Neurofibromas are an aberrant proliferation of Schwann cells, fibroblasts and perineural cells found throughout the body and are commonly associated with Neurofibromatosis-1 (NF-1) and Neurofibromatosis-2 (NF-2) [1]. Pediatric LNFs by comparison are quite rare with conflicting numbers reported in the literature [2–4]. Prior to 1990, the majority of patients with LNF were treated with open surgical resection. Recently there has been a trend toward the use of minimally invasive endoscopic techniques. In this case report we present an 8-year old boy with NF-1 who presented with a large asymptomatic supraglottic mass found on direct laryngoscopy during intubation for an unrelated procedure and will discuss the management strategies through a comprehensive review of the world literature.

2. Case report

An 8-year-old boy with NF-1 presented for evaluation of a supraglottic mass identified during intubation for an unrelated procedure. He had no history of airway compromise and was asymptomatic without dyspnea, dysphagia, or dysphonia. Physical examination revealed no stridor, retractions, or hoarseness. Flexible laryngoscopy revealed a submucosal mass centered within the left aryepiglottic fold extending inferiorly to the laryngeal ventricle and the medial wall of the piriform sinus (Fig. 1A). A CT scan of the head and neck revealed a well-circumscribed low-attenuated mass without contrast enhancement; axial and coronal MRI demonstrated contrast enhancement on T1-weighted images (Fig. 1B and C).

Given the concern for progressive airway obstruction based on the size and location of the mass, the patient was taken to the operating room for direct laryngoscopy and transoral CO_2 laser resection. Submucosal dissection exposed the tumor allowing gradual reduction of the tumor mass. Resection included the false fold mucosa in continuity with the underlying tumor. Dissection limits included the true vocal fold inferiorly, piriform mucosa posterolaterally, inner perichondrium of thyroid cartilage anterolaterally, and pharyngoepiglottic fold superiorly. Gross tumor resection was performed to clear gross but not microscopic disease in an attempt to limit post-operative functional deficits. The patient was extubated and transferred to the pediatric intensive care unit for 2 days for post-operative monitoring. A swallow study

* Corresponding author at: Department of Otolaryngology – Head and Neck Surgery; 1500 E. Medical Center Dr., Ann Arbor, MI 48109, United States. Tel.: +1 734 936 9598; fax: +1 734 936 4934.
E-mail address: mthorne@med.umich.edu (M.C. Thorne).
[1] Department of Otolaryngology – Head and Neck Surgery, University of Cincinnati, Cincinnati, OH, United States.

0165-5876/$ – see front matter © 2013 Elsevier Ireland Ltd. All rights reserved.
http://dx.doi.org/10.1016/j.ijporl.2013.10.047

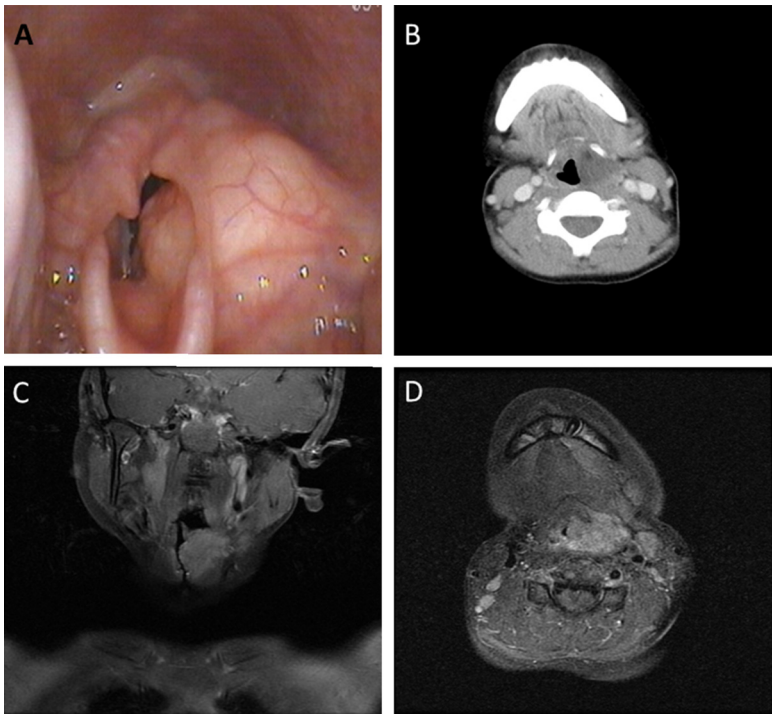

Fig. 1. (A) Pre-operative office-based flexible laryngoscopy demonstrating a large submucosal mass centered within left aryepiglottic fold with inferior extension to the false fold and lateral obliteration of the adjacent piriform sinus. (B) Axial CT-scan with contrast. (C) Representative coronal. (D) Axial MRI images.

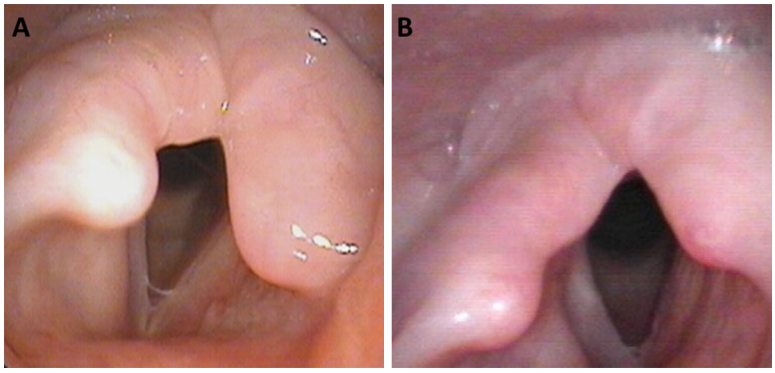

Fig. 2. Post-operative laryngoscopy: (A) 3-month and (B) 6-month.

was performed on post-operative day 1 revealing no aspiration and a normal pharyngeal phase swallow. An oral diet was started on post-operative day 1 and advanced without significant complications or clinical evidence of aspiration. Patient was discharged home on post-operative day 5 after an unremarkable hospital course. Pathologic analysis of the mass confirmed the diagnosis of LNF (Fig. 2). The patient was seen in routine follow-up at multiple intervals over an 18-month post-operative period without evidence of recurrence or additional symptoms. Fig. 3 shows the post-operative endoscopy at 3-months (left) and 6-months (right).

3. Discussion

Neurofibromas are peripheral nerve sheath tumors derived from peripheral nerves. Neurofibromas are associated with NF-1 and NF-2 or as spontaneous solitary lesions [1]. Neurofibromatosis-1 was first described by von Recklinghausen in 1882 [5]. The disease is an autosomal dominant disorder, although 30–50% of cases are associated with spontaneous germ-line mutations. Neurofibromatosis-1 typically presents in childhood and diagnostic criteria are defined as presentation with 2 or more of the

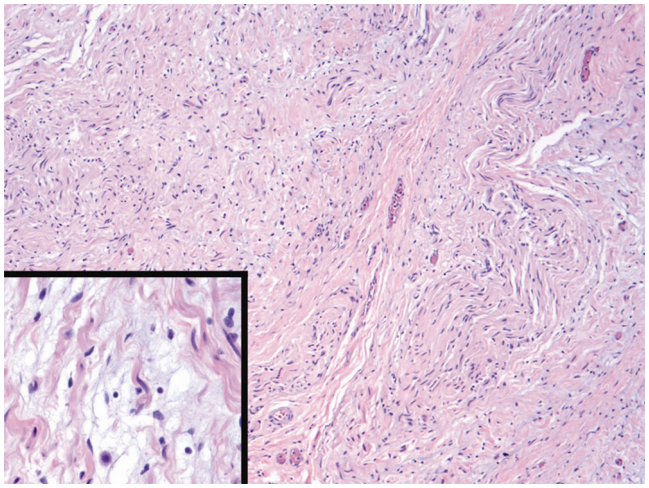

Fig. 3. Histology: the supraglottic biopsy consists of bundles of spindle cells set within a loose myxoid to collagenous stroma. At high power (inset), the spindle cells are associated with coarse collagen bundles and have hyperchromatic buckled nuclei characteristic of neurofibroma. (Hematoxylin and eosin 100×; inset 400×).