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Pediatric Laryngeal Neurofibroma: Case Report and Review of the Literature

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Abstract

Objective—Presentation of a case of pediatric laryngeal neurofibroma (LNF) and review of the world literature.

Data Sources/Review methods—Comprehensive review of the world literature using Pubmed and Google Scholar.

Results—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.

Conclusion—Pediatric LNF is a rare disorder. Review of the world literature since 1940 suggests a recent trend away from aggressive open resection and towards more conservative endoscopic resection with excellent functional results.

Keywords

Laryngeal neurofibroma; Neurofibromatosis; pediatric; transoral CO2 laser resection

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 towards the use of minimally invasive endoscopic techniques. In this case report we present an 8-year old boy with NF-1 who

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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.

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 (Figure 1, A). 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 (Figure 1B & 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 CO₂ 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 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 (Figure 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. Figure 3 shows the post-operative endoscopy at 3-months (left) and 6-months (right).

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 following: café-au-lait spots, neurofibromas, axillary/groin freckling, optic glioma, >2 Lisch nodules, sphenoid or long bone dysplasia, or first-degree relative with NF-1. Prevalence of NF-1 is cited in the literature as 1 in 2500 to 3000 patients. Neurofibromatosis-2 is an even rarer syndrome occurring in 1:40,000 live births with presentation typically in the third decade of life. Like NF-1, it is associated with neurofibromas and transmitted in an autosomal dominant fashion.

Bilateral acoustic neuromas are pathognomonic for NF-2, as well as the extra-cranial neurofibromas.^{2,6,7}

Laryngeal neurofibromas are an exceedingly rare tumor in the pediatric patient population with only 62 reported in the world literature (Table 1). Chevalier Jackson was the first to describe LNF while Colledge in 1930 was the first to describe LNF associated with neurofibromatosis.^{2,8} In 1940, Hoover described the first LNF in a pediatric patient.⁹ Chang-Lo et al. reviewed the worldwide literature in 1975 and identified 19 pediatric cases and in 1996, Masip et al. re-reviewed the world literature and identified 21 pediatric cases and until now represented the largest review of literature for pediatric LNF.^{2,4} This comprehensive review of the world literature in 2012 demonstrates that there have been 62 cases of pediatric LNF identified in case reports and case series. Table 1 lists all identified case reports and series published since 1940.^{1–46} There was an equal distribution of male to female cases (49.9% v 50.1%). The average age at presentation is 4.1 years (Range 0.8 – 12 years). In reports where NF-1 status was recorded, 82% (45/55) were associated with a diagnosis of NF-1 while only 18% (10/45) were identified in the absence of a NF-1 diagnosis. Review of presenting symptoms demonstrated stridor to be the most common presenting symptom (44%) followed by dysphagia (15%) and dysphonia (12%) (Figure 4B). Only 4%, including our index case, were identified incidentally.

From a diagnostic standpoint, flexible laryngoscopic examination demonstrates a highly vascular smooth submucosal mass most commonly in the supraglottis (Figure 1A). Terminal superior laryngeal nerves occupying the supraglottic submucosal space may explain the propensity for this anatomic subsite. As previously reported, the aryepiglottic fold is the most commonly involved subsite.⁷ Review of the reported anatomic distribution demonstrated the aryepiglottic fold was the most commonly cited location (34%) followed by the arytenoid (18%), and the post-cricoid/hypopharynx (14%). (Figure 4A) Preoperative imaging determines the lesion's anatomic extent, aiding in the selection of appropriate surgical approach. Unenhanced CT demonstrates hypo- to slightly hyperattenuated lesions; T1-weighted MR images demonstrate iso- to hyperintense lesions; T2-weighted MR images exhibit hyperintense lesions.³³ Variable enhancement is seen on both enhanced CT and MR images. (Figure 1B–D)

Surgical resection is the management of choice. Significant airway compromise will often require tracheotomy while non-obstructive lesions require resection to prevent airway, voice or swallowing sequelae of tumor growth over time. Review of treatment modalities prior to 1990 demonstrates that all reported cases required open surgical extirpation with the most extreme case requiring total laryngectomy in a 5 year-old female.³² Bagwell in 1990 was the first to report an endoscopic removal of a LNF with a CO2 laser.¹⁰ Since 1990, 30 cases of LNF have been reported and in cases where resection technique was described, 52% (12/23) have utilized endoscopic CO2 laser excision either as the sole modality or coupled with open surgical procedures. Tracheostomy has only been utilized in 33% (4/12) of cases using CO2 laser excision. Comparison to the pre-CO2 laser cohort is difficult given poor operative documentation, however in cases with detailed surgical reports, 66% (22/33) utilized tracheostomy. This suggests a diminished need for tracheostomy with an endoscopic approach.

The typical infiltrative growth pattern of neurofibromas often results in poor margin control and a proclivity for recurrence, even with aggressive surgical resection. Literature review demonstrated recurrence as a scarcely reported variable; however average follow-up was 4.9 years (median 4 years, range 0.5 – 15 years). With serial outpatient endoscopic surveillance, tumor recurrence can be identified and safely managed with repeat endoscopic resection as needed. Despite the tendency for recurrence, we favor a minimally invasive endoscopic approach over open techniques to limit postoperative morbidity associated with aggressive resection of laryngeal structures.

Although the incidence of LNF in NF-1 patients is low, any patient with NF-1 who presents with airway symptoms, voice changes or swallowing difficulty warrants further evaluation. We report an asymptomatic patient with LNF, demonstrating the need for a low threshold for work-up in this group of patients. Based on the presentation and subsequent follow-up of our patient; in conjunction with review of the world literature, endoscopic CO2 laser extirpation is a safe initial modality that may help avoid the need for open surgery. Additionally, minimally invasive techniques appear to show decreased need for tracheostomy in these selected patients.

Pitfalls of the current study are based on inherent biases of retrospective analysis and case report review. Given the retrospective case based literature review and the rare nature of this disease, absolute evidence based declarations of treatment cannot be made. Additionally prospective studies or comprehensive retrospective studies would be challenging if not impossible to produce given the rarity of this entity. Comprehensive reports in the literature at this time are the main mechanism for cursory evaluation of diagnostic and treatment efficacy.

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Figure 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 and (D) axial MRI images.

Figure 2.

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 100x; inset 400x).

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

Figure 4.

(A) Presenting symptoms and (B) anatomic distribution.

NOS = not otherwise specified; TVF = true vocal folds; AEF = aryepiglottic fold; HP = hypopharynx.

Table 1

Case reports and case series of pediatric LNF since 1940.

Reference (Year)	Age	Sex	Symptoms	Syndrome	Pathology	Location	Treatment	Follow-up
Hoover (1940) ⁹	10yrs	F	Dysphagia	NR	NF	False cord, AEF	Tracheostomy	6yrs, died
Smith (1944) ⁸	6yrs	M	Dyspnea on exertion	NR	NR	AEF	Tracheostomy, external approach without laryngeal violation	NR
	6yrs	M	Wheezing, stridor, cough	NR	NR	False cord, AEF	Tracheostomy, laryngofissure approach	NR
New (1947) ⁴⁶	5yrs	NR	Noisy breathing	NR	NR	Posterior surface of arytenoid cartilage	Thyrotomy approach	NR
Helg (1950) ²⁷	7yrs	F	Dysphagia, dysphonia	NF-1	NF	False cord, AEF	Laryngofissure approach	NR
Figi (1953) ⁸	5yrs	M	Stridor	No	NF	Posterior surface of arytenoids	Tracheostomy, lateral pharyngotomy	None
Mikell (1954) ³²	5yrs	F	Respiratory distress	No	NF	AEF, arytenoid	Lateral pharyngotomy, total laryngectomy	3yrs
Pung (1955) ³⁷	3yrs	F	Stridor	No	NF	Arytenoid, Posterior commissure	Tracheostomy, died before definitive treatment	NA
Gibbs (1957) ²¹	16mo	M	Stridor	NF-1	Plexiform NF	Postcricoid, trachea	Tracheostomy, died before definitive treatment	NA
Claux (1965) ¹³	2yrs	F	Stridor	NF-1	NF	AEF	NR	NR
Smoler (1966) ³⁹	2yrs	F	Stridor, dyspnea	NF-1	Plexiform NF	False cord	Tracheostomy, lateral pharyngo-laryngotomy	NR
Pleasure (1967) ³⁵	6mo	F	Stridor, respiratory distress	NF-1	Plexiform NF	Piriform fossa, AEF	Died before definitive treatment	NA
Goonewardene (1967) ²³	6yrs	M	Stridor	No	NF	AEF, arytenoids	Tracheostomy	NA
Gignoux (1968) ²²	10yrs	M	Dysphonia	NF-1	Plexiform NF	AEF	NR	NR
Johnsen (1970) ²⁹	11mo	F	Stridor, dysphagia	NF-1	Plexiform NF	Piriform fossa, AEF	Tracheostomy, neck exploration, pharyngotomy	1yr
Zagarskikh (1971) ⁴⁵	1yr	NR	NR	NR	NR	NR	NR	NR
Morris (1973) ²	18mo	M	Stridor	NF-1	Plexiform NF	AEF	ND	ND
Maisel (1974) ³⁰	11yrs	F	Stridor	NF-1	Plexiform NF	Epiglottitis, piriform fossa	Tracheostomy, pharyngotomy, neck exploration	3yrs
Chang-Lo (1977) ²	5yrs	F	Dyspnea, dysphagia, dysphonia	NF-1	Plexiform NF	Arytenoid, post-cricoid	Neck dissection, pharyngotomy	5yrs
Turchi (1977) ⁴²	3.5yrs	NR	NR	NR		NR		

Reference (Year)	Age	Sex	Symptoms	Syndrome	Pathology	Location	Treatment	Follow-up
Cohen (1978) ¹⁴	3yrs	F	Respiratory distress	NF-1	NR	False cord	Tracheostomy, laryngofissure	4yrs**
O'Connor (1980) ³⁴	7d	F	Stridor	NF-1	Plexiform NF	AEF	Pharyngotomy	3wks ^{##}
Garabedian (1987) ¹⁹	4mo	F	Dysphagia	NF-1	Plexiform NF	Piriform fossa, arytenoid, AEF	Partial laryngectomy	NR
	4yrs	M	Stridor	No	Plexiform NF	Epiglottitis, AEF, arytenoids	Partial laryngectomy	NR
Sidman (1987) ³⁸	2yrs	M	Dysphagia, dysphonia	NF-1	Plexiform NF	Arytenoid, Posterior false cord	Laser excision, tracheostomy	Multiple recurrence
Fukuda (1987) ⁷	2yrs	M	Stridor	No	NF	Subglottic	Tracheostomy, laryngofissure	NR ^{##}
Stanley (1987) ⁴¹	5yrs	M	Snoring	No	Plexiform NF	Arytenoid	Tracheostomy, thyrotomy excision	None
	2yrs	M	Stridor	No	NF	Subglottic, posterior larynx	Tracheostomy, thyrotomy excision	7yrs
Garashchenko (1988) ²⁰	NR	NR	NR	NF-1	NR	NR	NR	NR
Soboczki (1989) ⁴⁰	2.5yrs	M	Dyspnea	NF-1	NR	NR	NR	NR
Bagwell (1990) ¹⁰	NR	NR	NR	NR	NR	Larynx, not otherwise specified	CO2 laser	NR
Gupta (1991) ²⁶	10yrs	M	Stridor	NF-1	NF	Anterior commissure, TVF	Surgical excision, NOS	NR
Yamada (1992) ⁴⁴	7mo	M	Dysphonia, respiratory distress	NF-1	Plexiform NF	Post-cricoid, trachea	Died prior to surgery	NA
Martin (1995) ³¹	3.5yrs	M	Stridor	NF-1	Plexiform NF	Supraglottic larynx	NR	NR
	4yrs	F	Stridor	NF-1	Plexiform NF	Larynx, NOS	NR	NR
	6.5yrs	M	Stridor	NF-1	Plexiform NF	Larynx, NOS	NR	NR
	7 yrs	M	Asymptomatic	NF-1	Plexiform NF	Larynx, NOS	NR	NR
Gras Albert (1993) ²⁴	3yrs	M	Dysphonia	NF-1	Plexiform NF	NR	NR	NR
Willcox (1993) ⁴³	13mo	M	Stridor	NF-1	Plexiform NF	AEF	CO2 laser excision (x2)	1yr ^{∞∞}
Czigner (1994) ¹⁵	8yrs	M	Dyspnea, dysphonia	NF-1	NF	Piriform sinus, AEF, arytenoids	Tracheostomy, pharyngotomy	6yrs
Masip (1996) ⁴	4yrs	M	Stridor, dyspnea	NF-1	Plexiform NF	AEF, post-cricoid	Surgical excision, NOS	NR

Reference (Year)	Age	Sex	Symptoms	Syndrome	Pathology	Location	Treatment	Follow-up
Greinwald (1996) ²⁵	4yrs	F	Incidental Finding	NF-1	Plexiform NF	AEF, arytenoid	Partial supraglottic laryngectomy	NR
	7yrs	F	Stridor, dysphagia, dysphonia	NF-1	Plexiform NF	Post-cricoid, hypopharynx	Partial pharyngeal resection	2yrs
	9 yrs	F	Stridor, dysphagia	NF-1	Plexiform NF	AEF, arytenoid, piriform fossa	CO2 laser (multiple attempts), partial pharyngeal resection	1yr
	5mo	F	Stridor, dysphagia	NF-1	Plexiform NF	AEF, preepiglottic space	Partial pharyngeal resection, CO2 laser (x2)	14yrs
Pulli (1997) ³⁶	12yrs	F	Wheezing, dyspnea	No	NF	Subglottis	CO2 laser	5mo
Garabedian (1999) ¹⁸	6yrs	M	Stridor, dyspnea	NF-1	NF	AEF, false cord	Tracheostomy, lateral pharyngotomy, supraglottic hemi-pharyngotomy	1yr
	4yrs	M	Respiratory distress	NF-1	Plexiform NF	AEF, arytenoids, posterior commissure	Tracheostomy, Lateral pharyngotomy, supraglottic hemi-pharyngotomy, hemi-arytenoidectomy	9.5yrs
	2mo	F	Stridor, dyspnea	NF-1	Plexiform NF	AEF	Lateral pharyngotomy, supraglottic hemi-pharyngotomy	5yrs
	2yrs	F	Stridor	NF-1	Plexiform NF	AEF	Tracheostomy, Lateral pharyngotomy, supraglottic hemi-pharyngotomy	1yr
Chen (2002) ¹¹	18mo	M	Dyspnea, stridor	NF-1	Plexiform NF	AEF, false cord, posterior commissure, post-cricoid	Tracheostomy, Lateral pharyngotomy, supraglottic hemi-pharyngotomy	3yrs
	10yrs	F	Dyspnea	NF-1	NF	Piriform fossa, vallecula	Observation	NR
	8mo	F	Stridor, dysphagia	NF-1	NR	AEF	CO2 laser	4yrs
	6yrs	F	Dyspnea	NF-1	Plexiform NF	AEF	Endoscopic excision	NR
Rahbar (2004) ⁶	Birth	F	Stridor	NF-1	NF	Arytenoid, hypopharynx, cervical esophagus	Tracheostomy, partial pharyngeal resection	10yrs
	15w	M	Stridor, dysphagia, dysphonia	NF-1	NF	Neck, arytenoid, post-cricoid	CO2 laser	1yr
	1yr	F	Stridor	NF-1	NF	Pharynx, AEF, arytenoid	Tracheostomy, CO2 laser (multiple), supraglottic-laryngectomy, neck dissection, CO2 laser (x2)	15yrs
	10d	F	Stridor	NF-1	NF	Posterior commissure, arytenoids, mediastinum	Tracheostomy, CO2 laser (x2), neck dissection	5yrs
Chen (2004) ¹²	Birth	F	Stridor, dysphagia, dysphonia	NF-1	NF	AEF, arytenoid	Tracheostomy, CO2 laser (x4), endoscopic hemilaryngectomy	7yrs
	4yrs	F	Stridor	No	NF	AEF	CO2 laser	4yrs
	17mo	M	Dyspnea	NF-1	NF	AEF, posterior commissure	Tracheostomy, CO2 laser	6mo

Reference (Year)	Age	Sex	Symptoms	Syndrome	Pathology	Location	Treatment	Follow-up
Present Case (2009)	9yrs	M	Incidental	NF-1	NF	AEF, arytenoids	CO2 laser (x1)	1.5yrs

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NF = Neurofibromatosis; M = Males, F = Females; DOE = dyspnea on exertion; NR = Not recorded; NA = Not applicable; ND = Not Documented; TVF = true vocal fold; AEF = aryepiglottic fold; NOS = Not otherwise specified

** recurred 7 years after 1st extirpation, followed 4 years without recurrence.

Recurrence and Death 3 weeks after initial extirpation

∞∞∞ Recurrence 13 months after 1st extirpation, 2nd surgery curative.

∅∅∅ No Recurrence, but follow-up not noted.