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Isolated laryngeal lymphangioma: a rare cause of airway obstruction in infants

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Abstract

Laryngeal lymphangiomas are uncommon and virtually always represent extension of cervical disease. The exceptionally rare lymphangioma that remains isolated to the larynx has been reported only twice prior to this case.

A six-month male infant presented with an undiagnosed mass in the supraglottis which had caused respiratory compromise requiring a tracheostomy. The diagnosis was made histologically and the treatment was begun. Therapy consisted of staged laser resections carried out cautiously to preserve laryngeal competence. After three laser treatments the lesion was controlled and the patient prepared for decannulation. This interesting case is presented with a review of the literature.

Key words: Laryngeal neoplasms; Lymphangioma; Laser surgery; Child

Introduction

Lymphangiomas are rare congenital lesions of the lymphatic system which most commonly present in the head and neck during infancy (Emery *et al.*, 1984; Schloss *et al.*, 1984; Brock *et al.*, 1987). The larynx is a relatively uncommonly affected organ (Cohen and Thompson, 1986), the majority of head and neck lesions presenting with masses in the anterior and posterior cervical regions with variable extension into deeper tissues (cheek, tongue, floor of mouth, mediastinum) (Emery *et al.*, 1984). Laryngeal sites are usually identified in continuity with coexistent foci of disease in the neck with a reported incidence in a children's series of head and neck lymphangiomas of 14 to 22 per cent (Myer and Bratcher, 1983; Emery *et al.*, 1984; Cohen and Thompson, 1986). Cases of lymphangioma arising from the larynx and spreading to the neck have also been described (Williams and Cole, 1993). The rarest form of the disease, that in which the lymphangioma remains confined to the larynx has been reported only twice before in the English literature (Holinger and Johonston, 1951; Ruben *et al.*, 1975).

We present a case of lymphangioma arising in the supraglottis and remaining isolated to this single site. The method of management is described for this exceedingly rare laryngeal lesion of infancy.

Case report

A six-month-old male child (twin II) was referred for assessment of a laryngeal mass. The child had been evaluated at the referring institution for increasing stridor which had developed over the preceding three months. The stridor developed after an upper respiratory tract infection, was constant and showed no alteration during sleep or with activity. The child had difficulty feeding compared to his twin although his mother did not think he

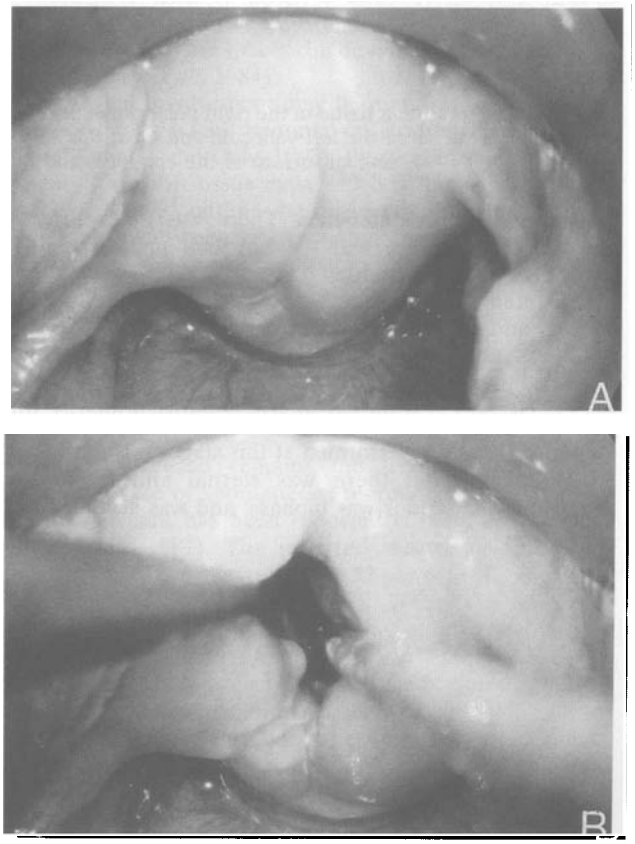

FIG. 1

A: Endoscopic appearance of the lymphangioma confined to the supraglottis. B: The glottis is normal but can only be viewed if the supraglottis structures are pried apart with the probe and the suction

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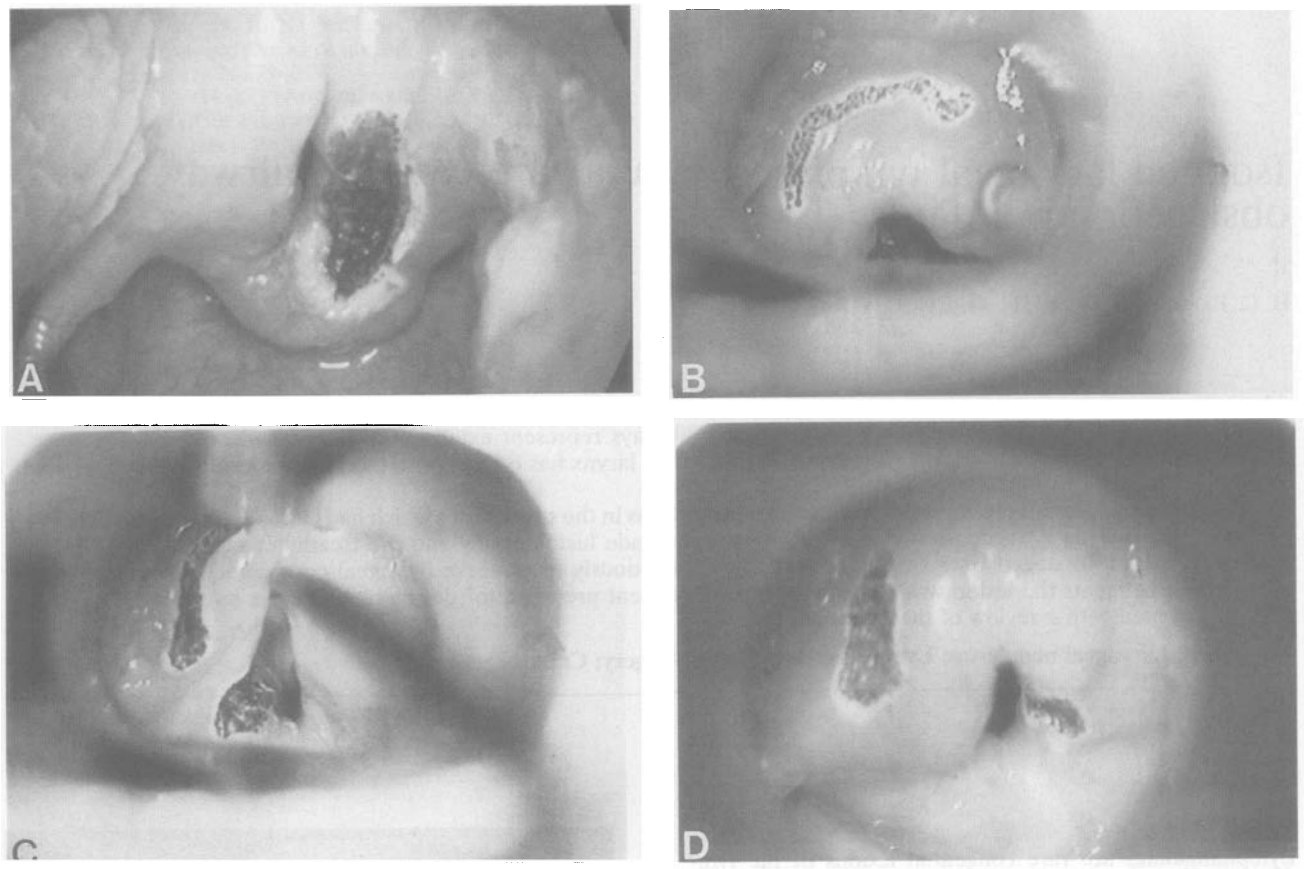

FIG. 2

A: At the first treatment tissue in the right posterior supraglottis was resected with the laser. B: The subsequent treatment involved resection of tissue from the left vallecula and C: at the base of the left aryepiglottic fold. D: At the final laser treatment the left lingual surface of the epiglottis and the right posterior aryepiglottic fold were lased.

was limited by breathlessness. There was no history of aspiration or choking and the boy had maintained his growth on the third centile.

The infant was born at 37 weeks gestation by forceps delivery and had Apgars of 9 at one and five minutes. The child was admitted to the special care baby unit and remained there because of poor feeding for two and a half weeks. He was readmitted at five months primarily for poor feeding but was also noted at this time to have stridor. A barium swallow performed at this stage was normal.

On examination there was sternal and intercostal recession. The stridor was biphasic and was accompanied by a tracheal tug. There was good air entry bilaterally and evidence of slight hyperinflation. The liver was palpable at 1.5 cm below the costal margin, this might also have been a manifestation of pulmonary hyperinflation. A 1/6 systolic murmur at the left sternal edge was felt to represent a flow murmur. The clinical impression at the referring institution was that the child had laryngomalacia and the child was scheduled for an endoscopic examination. At endoscopy, the epiglottis was swollen and the swelling continued laterally onto the right aryepiglottic fold (Figure 1A). The vocal folds and arytenoids were normal (Figure 1B) but could only be viewed after the probe and suction were used to open the supraglottis. A tracheostomy was performed and subsequently transfer was arranged to our institution.

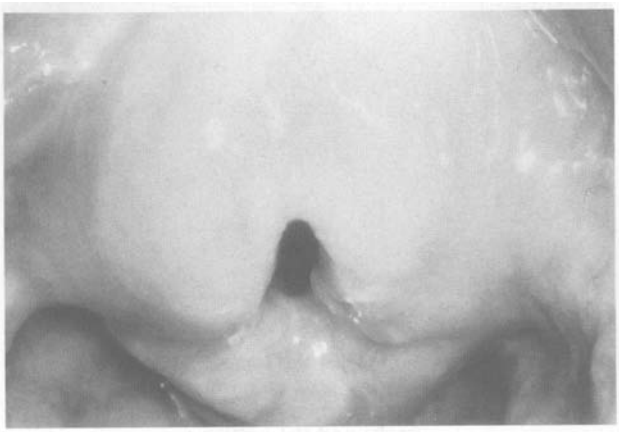

FIG. 3

Two months after completion of the laser resections the larynx is competent and the child ready for decannulation.

Upon arrival the airway was secure. An ultrasound of the neck revealed no collection or laryngeal cyst and a soft-tissue radiograph of the neck showed a swollen supraglottic larynx. Re-examination of the larynx under general anaesthesia confirmed findings identical to those described prior to transfer. Some oedematous mucosa was taken from the left aryepiglottic fold and sent for histological examination and some swabs were taken and sent for bacterial and viral culture. No significant pathogens were isolated. A pH probe study was arranged to exclude reflux noting that a barium swallow had previously been reported as normal. The result showed moderate gastrooesophageal reflux 120 minutes post-prandially.

Histopathology revealed squamous epithelium with a focal inflammatory infiltrate. In the underlying connective tissue there was a widespread proliferation of thin walled vessels with fine, partial septae or valve-like structures.

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TABLE I

INCIDENCE OF LARYNGEAL LYMPHANGIOMA IN SERIES CONFINED TO LYMPHANGIOMAS OF THE HEAD AND NECK AND BENIGN LESIONS OF THE LARYNX

Author	Year	Number of patients in series	Number of laryngeal lymphangiomas	Lymphangiomas included in series of-
A. Bill and Sumner**	1965	61	1	lymphangiomas—head and neck
Cohen and Thomson**	1986	73	10	lymphangiomas—head and neck
Emery *et al.***	1987	37	8	lymphangiomas—head and neck
B. New and Erich	1938	722	1	benign laryngeal lesions
Holinger and Johonston	1951	1197	1*	benign laryngeal lesions
Anderson**	1951	768	48	benign laryngeal lesions
Holinger *et al.*	1965	866	6	benign laryngeal lesions

*Case of lymphangioma confined to the larynx.
**Exclusively paediatric series.

The diagnosis of lymphangioma was made. The patient remained well but was returned to the operating-theatre one month later for treatment. His weight gain had improved. There was still no evidence of any masses in the neck.

The plan for treatment was to perform conservative excisions moving to different sites within the supraglottis at each treatment. Minimal excisions were carried out at each session and care was taken to minimise ablation so that the larynx would remain functionally competent. At the first treatment the CO_2 laser (5 Watts continuous) was used to vaporize tissue in the right posterior glottis (Figure 2A). The child overspilled for four days post-operatively and remained in hospital during this time. One month later he returned to the operating-theatre and the laser was used to resect tissue from the left vallecula (Figure 2B) and at the base of the left aryepiglottic fold (Figure 2C). The laryngeal introitus was still not open. The resection was purposely minimised to avoid post-operative overspilling and none occurred. Two months later, another laser treatment was performed with lasering carried out to the left lingual surface of the epiglottis and the right posterior aryepiglottic fold (Figure 2D). The larynx was now minimally opened. At the next endoscopic evaluation two months later the larynx was competent (Figure 3) and there was no evidence of aspiration. The child was successfully decannulated eight months after his initial tracheostomy. There has been no sign of any additional sites of lymphangioma nor has there been any continguous spread of the lesion within the neck. This represents an exceptionally rare case of lymphangioma presenting solely as an airway-compromising laryngeal lesion.

Discussion

Lymphangiomas are rare congenital lesions of the lymphatic system which most commonly present in the head and neck during infancy (Emery *et al.*, 1984; Schloss *et al.*, 1984; Brock *et al.*, 1987). There has been a considerable amount of debate since the lesion was first described about the development of the lymphatic system and the developmental error which leads to formation of lymphangiomas. Some theorists have suggested that the lymphatic primordia grow outward (centrifugal theory) from venous channels (Groetsch, 1938) while others believe the lymphatic system develops independently of the veins (centripetal theory) and establishes connection with the venous system later (McClure and Sylvester, 1909). In either case, prominent lymph sacs including the bilateral cervical jugular sacs are detectable during the seventh week of embryonic life (Sabin, 1901). Whether these sacs fail to make contact with the venous system or fail to maintain their previously developed connections is a point best left disputed by the embyrologist. It is clear, no matter which developmental theory is held, that the pathological lesion results from lymphatic cysts isolated from their normal route of drainage into the venous system and they cause their effect by expansion following this sequestration. Expansion may result from the sequestered tissue's proliferative growth potential (McClure and Sylvester, 1909) or may follow infection or trauma to a previously quiescent cyst (Leipzig and Rabuzzi, 1978).

Histologically these lesions are comprised of normal mature lymphatic tissue which can be subdivided into capillary, cavernous and cystic types by the size of the lymphatic spaces. Commonly all three subtypes coexist within a single lesion (Batsakis, 1979). Cystic hygroma is a clinical term used to describe large cystic neck masses which occur in areas where expansion can occur and large multiloculated cystic spaces can develop, usually low in the neck (Emery *et al.*, 1984).

Laryngeal involvement may be manifest as hoarseness, dyspnoea, dysphagia, aphonia or stridor (Emery *et al.*, 1984; Williams and Cole, 1993). Physical examination may reveal a firm soft-tissue mass which is fluctuant. Williams and Cole (1993) outline the diagnostic imaging currently used in the assessment of lymphangiomas of the larynx and advocate the use of plain radiographs of the neck and chest to evaluate the extent of oesophageal and laryngeal obstruction. Ultrasound can be valuable in pre- and post-natal diagnosis and in monitoring the extent of the lesion. MRI delineates clearly the cystic mass from the surrounding normal tissue but CT is of value to differentiate the cystic lesion from vascular tumours and mucus retention cysts.

Surgical excision remains the treatment of choice although a rate of spontaneous regression between eight and 15 per cent has been reported (Emery *et al.*, 1984; Brock *et al.*, 1987). The reported recurrence rate from residual lesions ranges from 10 to 52 per cent (Ravitch and Rush, 1979; Emery *et al.*, 1984). When the larynx is involved tracheostomy is almost always required (Emery *et al.*, 1984; Cohen and Thompson, 1986). Endoscopy and repeated CO_2 laser excision remain the preferred method of managing the laryngeal foci and vaporization of the cyst walls in addition to deroofing the lesions is advocated to reduce local recurrence (Williams and Cole, 1993). Occasionally more extensive resections are required and modified radical neck dissections (Williams and Cole, 1993) and partial laryngectomy (Myer and Bratcher, 1983) have been used to control lesions in the neck with laryngeal involvement. The benign nature of lymphangiomas should be considered in treatment planning and preservation of neurovascular structures accomplished whenever possible (Myer and Bratcher, 1983; Emery *et al.*, 1984).

Lymphangiomas represent approximately six per cent of non-inflammatory masses of the head and neck in children

(Anderson, 1951). The rarity of laryngeal involvement in lymphangioma can be seen in Table I. In all but one of the cases* shown in Table I, the laryngeal lymphangioma represented extension of a cervical lesion. Williams and Cole (1993) report a case of an apparently isolated laryngeal lymphangioma in a neonate treated with CO_2 laser but the infant went on to develop extensive cervical extension of disease requiring tracheostomy and neck dissection. Isolated laryngeal lymphangioma, though exceptionally rare, has been previously reported.

Naito *et al.* (1985) reviewed the entire English literature and documented 17 cases in which there was isolated laryngeal involvement with lymphangioma. Notably, all but two occurred in adults. One of the paediatric lesions was located in the subglottis (Holinger and Johonston, 1951) and the other in the vallecula (Ruben *et al.*, 1975). The surgical details of the three-year-old female with a subglottic focus reported in 1951 are incomplete although it is clear a tracheostomy was performed (Holinger and Johonston, 1951). Ruben *et al.* resected a lymphangioma from the vallecula of a 20-month-old male on two occasions (one recurrence at 14 months) and the child did not require a tracheostomy (Ruben *et al.*, 1975). Our current case illustrates the value of this endoscopic laser resection in the treatment of this lesion. The selective ablation of affected tissue allowed careful recontouring of the larynx permitting preservation of competence and function.

Summary

Isolated laryngeal lymphangioma is an exceptionally rare cause of airway obstruction in infants. Establishment of a secure airway often with a tracheostomy, is the initial management priority. Surgical excision with preservation of vital structures remains the treatment of choice. The surgical laser is particularly well suited for treatment of this lesion as it allows precise ablation and recontouring of the laryngeal tissue so that functional competence can be preserved.

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