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Lymphatic malformations of the tongue base

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Debulking procedures;
Laryngeal involvement;
Airway complications

Abstract

Purpose: In light of the paucity of literature on lymphatic malformations of the tongue base, our aim was to present our experience and long-term outcomes of patients with this rare and challenging pathologic entity.

Methods: Medical records of 25 patients treated by the 3 senior authors (RGA, MTC, and RTC) between 1974 and 2003 were retrospectively reviewed, and comprehensive clinical data were collected and analyzed.

Results: Twenty-one patients (13 female and 12 male infants) were diagnosed either prenatally or at birth. Of these patients, 18 required early airway stabilization; 17 required tracheotomy. Four patients were diagnosed after 1 year of age and had no airway problems. Follow-up ranged from 2 days (owing to death) to 28 years, with a mean of 10 years. In 21 patients, pathology was extensive, involving contiguous anatomical areas such as the anterior tongue, larynx, pharynx, and floor of mouth. Multiple resections and debulking procedures were performed to restore function and improve cosmesis. Four patients died, all with laryngeal involvement. Of the 14 survivors who had tracheotomies, only 5 are decannulated. Normal oral feeding has been achieved in 14 patients and normal speech, in 8 patients. Cosmesis has improved with time. Orthodontic and dental problems are common, and 9 patients have significant macrognathia.

Conclusions: Although most patients with lymphatic malformations of the tongue base achieve normal oral feeding, airway, speech, and cosmesis issues remain problematic throughout life. Laryngeal involvement signifies extensive disease and is the most significant risk factor for serious complications and death.

Lymphatic malformations (lymphangiomas) appear to arise from embryological disturbances in the development of the lymphatic system. These abnormalities occur in various anatomical sites; however, more than 50% are located in the head and neck region [1]. Lymphatic malformations of the tongue are rare [2-4], with most cases affecting its anterior

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portion [1,5]. To date, published series and case reports [1-3,5-10] have not specifically addressed tongue base involvement and, thus, provide clinicians with little if any information on the management of these lesions. Although both anterior and tongue base involvement can lead to facial deformities, impairment of speech, and secondary psychological problems, the clinical profile associated with the latter is generally far more complex. The risk of laryngeal involvement, upper airway obstruction, and feeding problems is extremely high. Treating patients successfully requires a thorough understanding of head and neck anatomy, a broad surgical armamentarium, and comprehensive interdisciplinary team support.

To shed light on this rare and challenging anomaly, we present here our collective experience with 25 children diagnosed with lymphatic malformations of the tongue base (LMTB) over a 29-year period. Our objective was to gather information that could be used to identify key prognostic factors, to clarify the impact of specific therapeutic interventions, and to determine long-term outcomes.

1. Patients and methods

We combined the personal series of the 3 senior authors (RGA, MJR, and RTC). The medical records of patients they treated for LMTB between 1974 and 2003 were retrospectively reviewed. Collected data consisted of demographic information and a wide range of clinical parameters, including airway management, the extent of anatomic involvement, surgical interventions and other treatment modalities used over time, and effects on feeding, speech, and cosmesis. Because of insufficient data, 4 patients were excluded from the study. Approval for this project was granted by our institutional review board.

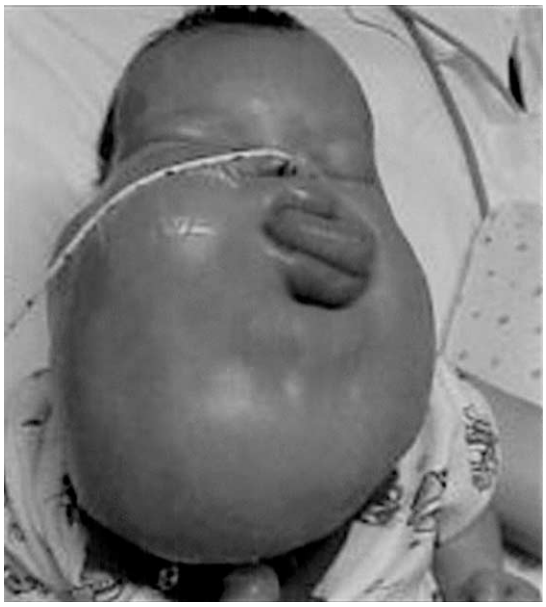

Fig. 1 Neonate with massive cervicofacial lymphatic malformation involving the tongue. This infant required a tracheostomy to secure the airway.

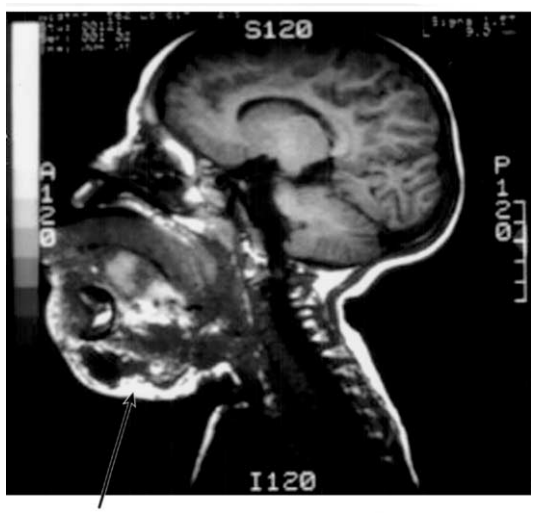

Fig. 2 Craniofacial magnetic resonance image of a child with an extensive cervical lymphatic malformation involving the tongue base and floor of mouth.

2. Results

The study population comprised 25 patients with LMTB. Sex distribution was almost equal, with 12 male and 13 female infants. Of 25 patients, 9 were diagnosed in utero, 12 were diagnosed at birth, and 4 were diagnosed later than 1 year of age. Follow-up ranged from 2 days (owing to death) to 28 years, with a mean of 10 years and a median of 6 years.

With the exception of 1 patient, all had bilateral base of tongue involvement (Fig. 1). None of the 4 patients who were diagnosed after 1 year of age required a tracheotomy. By contrast, all 9 patients diagnosed in utero required early airway intervention, with 3 requiring ex utero intrapartum treatment (EXIT) procedures,[1] and 8 requiring tracheotomies. Of the 12 patients diagnosed at birth, 9 required tracheotomy. Thus, a total of 18 patients required immediate airway intervention at birth. After airway stabilization, most patients received computed tomographic scans or magnetic resonance imaging to help evaluate the extent of disease (Fig. 2).

Extensive pathology was present in 21 of 25 patients, whereas localized disease was seen in only 4 patients; 3 of these 4 patients were diagnosed after 1 year of age. As

[1] EXIT procedure: this procedure maintains placental circulation to the fetus while securing the airway at the time of delivery. Instrumentation of the airway, including tracheostomy, may be accomplished at this time. Candidates for the EXIT procedure have airway obstruction secondary to large cervical masses.