Case Report

Emergent sclerotherapy of a newborn with expanding lymphatic malformation causing respiratory distress

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\textbf{ABSTRACT}

The present report describes a case of acute airway obstruction in a newborn caused by an expanding hemorrhagic macrocystic lymphatic malformation (LM), which was successfully treated with emergent decompression and interventional radiology-guided sclerotherapy. The use of sclerotherapy for macrocystic LMs has been well described for various indications. The urgent interventional treatment obviated the need for a tracheostomy. This case describes the rapid diagnosis and use of sclerotherapy in a large expanding macrocystic LM.

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\textbf{Introduction}

Lymphatic malformations (LMs) are benign abnormalities that typically present in the head and neck region as painless neck masses. Fifty percent of these lesions are noted to be at birth and 80%-90% of these lesions are diagnosed before the age of 2 years [1]. Although the etiology of LM is still unclear, recent studies have shown intrinsic and extrinsic factors to play a role, including inappropriate expression of lymphatic specific molecules and increased number of interferon producing cells [1–3]. LMs account for only 5% of all benign pediatric lesions and frequently involve the head and neck region. However, because of their variable clinical course, fluctuation in size, and mass effect on nearby structures, they may pose life threatening risks to this population [4].

We present a unique case of a rapidly enlarging macrocystic LM in a 17-day-old newborn that was thought to be due to hemorrhage. The patient underwent emergent airway
evaluation and intervention with resultant intubation and sclerotherapy. Intraoperative evaluation revealed a 5.3 × 4.2 × 3.3 cm cyst in posterior pharynx and deep neck deviating the trachea anteriorly. This case demonstrates intracystic hemorrhage in a newborn with an LM and describes successful decompression and sclerotherapy treatment obviating the use of a tracheostomy.

Methods

Our institution does not require approval for retrospective studies such as this. This retrospective case report includes clinical history, intraoperative findings, radiographic findings, and a review of the current literature.

Case presentation

A 17-day-old normal full-term female presented to the emergency room of a tertiary pediatric hospital with a 1-day history of acute onset respiratory distress and enlarging neck mass over the course of an hour. She had no significant medical history, and there was no reported history of trauma, foreign body aspiration, or previous intubations. On arrival, she demonstrated increased work of breathing, gasping with marked retractions, and cyanosis. She was placed on 100% nonrebreather mask and given racemic epinephrine. Jaw thrust with positioning improved oxygen saturations to the 90s. Urgent otolaryngology evaluation revealed an enlarging neck mass with impending airway obstruction, which prompted the decision to secure the airway in the operating room.

She underwent direct laryngoscopy, which revealed massive anterior retropharyngeal displacement effacing the laryngeal surface and obstruction of the airway. Her airway was secured with a 3-0 uncuffed endotracheal tube, and patient was then transferred to the computed tomography (CT) scanner to investigate developmental (thyroglossal duct cysts, branchial cleft cysts, etc), inflammatory and/or infectious, and neoplastic causes.

Contrast CT scan of the neck demonstrated a large bilobed cystic lesion measuring 5.3 cm × 4.2 cm × 3.3 cm displacing the trachea anteriorly with evidence of increased attenuation consistent with internal hemorrhage (Figs. 1 and 2). Her findings were consistent with a large macrocystic LM with acute hemorrhage; she was then transferred to intervention radiology to undergo sclerotherapy.

Using fluoroscopic guidance and ultrasound, a 5-French catheter was placed and subsequently aspirated 13 milliliters of bloody fluid. There was noted to be significant decompression of the cyst with resolution of mass effect. The cavity was irrigated and chemical thrombolysis was performed on the remaining clot, which was completely aspirated. Sclerotherapy was then performed with 5 milliliters of alcohol with subsequent aspiration of 6 milliliters of bloody fluid (Fig. 3). The drain was secured in place and patient was taken back to the operating room for further evaluation of her airway.

Repeat direct laryngoscopy revealed resolution of mass effect with slight retropharyngeal fullness and an uncompromised view of the larynx. The 3-0 uncuffed tube was replaced with a 3-0 cuffed tube without difficulty. Bronchoscopy showed no evidence of tracheomalacia. After completion of the procedure, she was placed on prophylactic cefazolin and transferred to the Pediatric Intensive Care Unit for further monitoring.

The patient continued to improve through her hospital course. The 5-French drain was removed on postoperative day three (POD 3). She was subsequently weaned from the ventilator and after bedside direct laryngoscopy revealed

Fig. 1 – Computed tomography (CT) scan axial view demonstrating macrocystic malformation.

Fig. 2 – CT scan coronal view demonstrating compression of the airway.
no pharyngeal or laryngeal obstruction, patient was extubated on POD 4. She had no evidence of stridor or respiratory distress and was transferred to the floor on POD 5. Nasopharyngeal fiberoptic scope examination on POD 6 revealed slight nonobstructive retropharyngeal asymmetry, thereby prompting ultrasound evaluation. Ultrasound revealed a collapsed cyst around a central blood clot and solid intracystic nodule, requiring no further intervention at that time. Patient continued to do well and was discharged home on POD 8 with pulse oximeter for continued surveillance.

She returned for office evaluation on POD 13 and physical examination was notable for 1 cm × 2 cm firm left cervical mass. Nasopharyngeal scope displayed left pharyngeal asymmetry with no signs of respiratory compromise or obstruction. This was followed by ultrasound evaluation, which revealed a 2.3 cm × 1.2 cm bilobed cystic lesion for which she received additional sclerotherapy treatment. It was noted at that time that there were 4 other small microcystic lesions, all of which were subsequently treated in a similar manner. Interventional radiology follow-up 2 months after the initial procedure displayed few tiny microcystic lesions and resolution of the large macrocystic lesion.

At her most recent follow-up, patient had not had any episodes of respiratory difficulty. She underwent magnetic resonance imaging evaluation demonstrating small reactive posterior cervical lymph nodes bilaterally with no dominant cystic lesion or enhancing soft tissue neck mass (Figs. 4 and 5). She continues to progress well from a developmental and respiratory standpoint and has had no recurrence of the LM for 3 years.

Discussion

The assessment and treatment of LMs has improved over the last 15 years because of simplified LM nomenclature, an LM staging system, detailed imaging with better characterization, and innovations in both sclerotherapy and surgery.
LMs are classified as macrocystic, microcystic, or mixed [1]. Macrocystic LMs consist of a single or multiple cyst > 2 cm³, whereas microcystic LMs are comprised of single or multiple cysts < 2 cm³. Mixed LMs have components of both macro- and microcystic lesions. LMs are staged in accordance with laterality and anatomical relationship of the lesion to the hyoid bone [5]. Although preoperative staging is necessary for treatment planning and prognostic discussion, we were unable to complete this because of the acuity of the patient's presentation. On later obtainment of imaging studies, we were able to characterize the lesion as a macrocystic LM.

Symptoms are related to the size of the LM and its altering effects on the structure and function of adjacent tissues. Sudden enlargement of these lesions may be due to recurrent inflammation, infection, hemorrhage, or trauma, thereby causing life threatening impairment of vital structures [3]. Although our case lacked antecedent trauma or infection, imaging identified acute hemorrhage into the cystic space causing mass effect and respiratory distress. Multimodal treatment is used to achieve favorable outcomes in this patient population, including surgery, sclerotherapy, or a combination of both. Notably, sclerotherapy has been shown to be effective in the treatment of macrocystic LMs and reduces the need for other forms of therapy [6–8]. Although preferred for being minimally invasive, complications such as skin blistering, nerve damage, and tissue necrosis have been noted in the literature. However, because of the acuity of the situation and difficulty of complete surgical resection, this treatment option was used in our case after an extensive multidisciplinary assessment.

Although LMs are known to cause airway compromise when key structures of the larynx and tongue are involved, few cases have been documented with definitive management at the initial presentation in an infant. Although many practitioners may opt for surgical management in acute cases with airway involvement including a tracheostomy, this report details successful management of a fully effaced airway in a neonate via decompression and sclerotherapy. This case details a management strategy for an obstructing LM with acute hemorrhage with the use of interventional radiology techniques in a collaborative manner with otolaryngology service without the concomitant use of a tracheostomy.

**Conclusion**

Differing presentations and characteristics of LMs have been well described in the literature. Less commonly described is the onset of acute respiratory distress in the neonatal population secondary to macrocystic LMs. Given the paucity of similar cases available, management has not yet been standardized. We present a novel use of sclerotherapy in an emergent infant airway obstruction demonstrating the feasibility of management without a tracheostomy.

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