Childhood airway manifestations of lymphangioma: A case report

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Lymphangioma is a congenital malformation of the lymphatic system, often involving areas of the head and neck. The involved structures may include enlarged tongue and lips, swelling of the floor of the mouth, and direct involvement of the upper respiratory tract. The definitive treatment for lymphangioma is surgery, often during the first years of life. Despite surgical removal, lymphangioma may persist. Anesthetic concerns include bleeding, difficulty visualizing the airway, extrinsic and intrinsic pressure on the airway causing distortion, and enlarged upper respiratory structures, including the lips, tongue, and epiglottis.

This is a case report of a 9-year-old patient with lymphangioma who had impacted teeth and a suspected odontogenic cyst. There seems to be little information on the optimal anesthetic management for this age group. The challenges with airway management, including bleeding, laryngospasm, and a difficult intubation, are outlined.

Awareness of potential airway involvement and possible complications is necessary to provide a safe anesthetic to a patient with lymphangioma. A review of the literature, airway management techniques, and current airway equipment will be discussed.

Key words: **Airway management, anesthesia, intubation, lymphangioma.**

Lymphangioma is a congenital malformation of the lymphatic system. Cystic hygroma, a type of lymphangioma, is composed of large lymph-containing cysts.[1] These benign, painless, soft malformations affect both sexes equally.[2] They usually are found at birth, with 90% diagnosed before the age of 2 years.[3] Cystic hygroma can arise in any area of the body; however, more than 90% of lymphangiomas are in the cervical region.[1] The larynx, the orbit, the tongue, floor of the mouth, and the cheek also may be involved.[2] Airway complications, dysphagia, and speech difficulties may develop.[4] The most life-threatening complication of lymphangioma is acute airway obstruction.[5]

The treatment of lymphangioma is primarily surgical. Complete surgical excision is balanced with keeping vital structures intact.[6] Maintaining this balance may result in incomplete excision and, eventually, recurrence, especially when the hygroma involves the upper airway. The most common places for recurrent disease are lesions of the floor of the mouth, lip, tongue, and hypopharynx.[6] It is important to note that residual disease in the upper airway needs special consideration, even for surgical cases unrelated to the lymphangioma itself, with implications for airway management.

The following is a case report of a child with a previously excised lymphangioma admitted for dental surgery. Anesthetic considerations, including airway management, alternative airway equipment, and possible complications, are discussed.

Case summary

A 9-year-old girl with orofacial and cervical lymphangioma was admitted to have an odontectomy of 8 retained primary mandibular teeth. The patient also had a suspected eruption cyst in the area of teeth numbers 5 and 6. The patient's history included multiple surgeries directed toward management of extensive lymphangioma, with the most recent surgery occurring 6 years earlier.

The patient was delivered via cesarean section at 38 weeks of gestation due to notation of decreased fluid flow through the trachea as seen on ultrasound. The delivery was planned to allow intubation while still maintaining maternal-fetal circulation. Due to the massive lymphangioma with involvement of the oral cavity, oropharynx, and hypopharynx, rigid laryngoscopy and bronchoscopy were required to establish the airway with subsequent endotracheal intubation. The patient had extensive lymphangioma involvement, including the oral cavity, oropharynx, supraglottic larynx, anterior portion of the face, and both sides of the neck. The initial resection was undertaken at 2 days of age, addressing primarily the cervical component of the lymphangioma. Attempts at extu-

bation failed due to lymphangiomatous involvement of the oral cavity, oropharynx, and supraglottic larynx. A tracheostomy was performed at 27 days of age.

Multiple treatments addressing the residual lymphangioma involving the tongue base, supraglottic larynx, and oral cavity were completed. Further surgery addressing the extensive cervical facial component of the lymphangioma included a total parotidectomy and dissection of the submandibular and submental regions. At age 3 years, the tracheostomy was closed successfully, and a stable airway has been maintained since that time.

Since the closure of the tracheostomy, other health issues have included otitis media, lymphangiomymatosis with involvement of the right orbit, and intermittent episodes of pharyngitis. The patient was functioning at appropriate levels socially and was progressing normally in school.

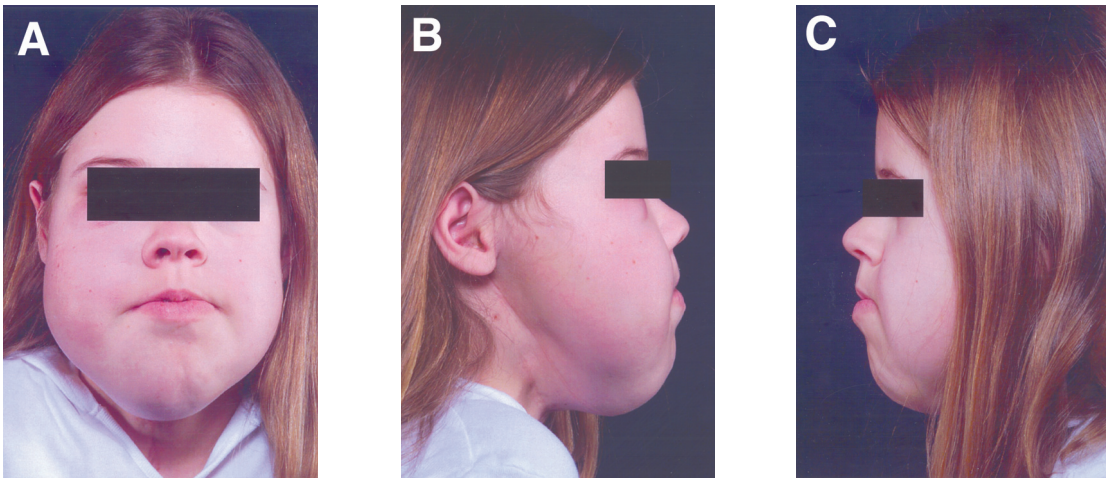
During the preanesthesia examination for the odontectomy, it was noted that the patient had a fullness of cheeks that extended down to the neck, mostly on the right side (Figure). The patient had no evidence of hypognathia and was able to open her mouth without difficulty. It was noted that the patient's speech was slightly dysarthric. The patient had been clear of respiratory and ear infections for 2 months, and her parents reported no evidence of airway obstruction. The patient was extremely cooperative. After consideration of an awake fiberoptic intubation, it was decided to proceed with a mask induction. The anesthetic plan of the mask induction was explained to the patient and parents.

On arrival to the operating room, routine monitors, including a pulse oximeter, noninvasive blood pressure cuff, and electrocardiographic monitor were applied. A mask induction with a nasal intubation was planned. The patient was preoxygenated with 100% oxygen for approximately 3 minutes. A precordial stethoscope was applied. Nitrous oxide in oxygen was added at 70%, and sevoflurane was added incrementally. The patient's respirations were assisted with positive pressure and a mask airway was maintained.

As the anesthetic was deepened, the tongue relaxed into the airway, causing a partial obstruction, and placement of a number 8 oral airway was attempted. The oral airway would not fit around the base of the tongue, which was noted to be hard with many coarse bumps and immediately started to bleed with only slight pressure from the oral airway. As the oral airway was removed, the patient experienced a laryngospasm. Positive pressure was administered via the facemask, while a 20-gauge intravenous catheter was inserted, and 30 mg of succinylcholine was given intravenously. The laryngospasm abated, and ventilation with 100% oxygen was continued. The patient's tongue continued bleeding, and oropharyngeal suctioning was instituted.

During the laryngoscopy, it was difficult to insert a Miller 2 blade because the tongue was hardened, especially in the posterior oropharynx. Oral intubation

Figure. Nine-year-old girl who underwent lymphangioma resection as an infant



(A) Note fullness of the cheeks extending to the eye area. (B) Note fullness of the cheeks and enlarged neck, predominantly on the right side. (C) Facial fullness is less predominant on the left side of the face.

required modest cricoid pressure for visualization of the slightly anterior vocal cords. Once the airway was established, further discussion with our surgical colleagues determined that the optimal surgical exposure would be obtained with nasopharyngeal intubation. When it was determined that the cords could be visualized, a number 5 endotracheal tube was passed through the patient's nose. Under direct visualization, the oral endotracheal tube was removed and the nasal tube was placed. It was noted that the inner lower lip, tongue, and side of the cheek were hardened with multiple coarse spots that were bleeding.

The patient's oxygen saturation stayed above 90% throughout induction and intubation. The patient's oxygen saturation and vital signs remained stable for the rest of the case. Six teeth were extracted, and a cyst near the number 5 tooth was enucleated. Three milligrams of ondansetron were given to abate postoperative nausea. Because of the tongue size and potential airway problems, it was decided that the patient would need to meet strict awake extubation criteria. A 26 French nasopharyngeal airway was placed in the opposite nostril; the patient's mouth and pharynx again were suctioned. When the patient appeared fully awake, followed all commands, and showed a strong head lift, extubation was accomplished successfully. During the postoperative period, the patient showed no signs of airway complications. Postoperative follow-up was done with the patient's parents to encourage the parents to obtain an airway Medic Alert bracelet for the patient.

Discussion

This case highlighted the potential anesthetic complications with patients undergoing intubation with residual oropharyngeal lymphangioma. Most medical literature has focused on the surgical and airway management of this malformation at the time of birth and through the first series of surgeries in early life. There seems to be little information on the optimal anesthetic airway management for this age group.

There are several characteristics of lymphangioma that need to be anticipated when providing anesthesia. The first concern is the extent to which the lymphangioma has affected the airway. Anatomical airway changes resulting from the staged series of surgeries for cystic hygroma in the head and neck area are difficult to assess. As in all potentially difficult airway cases, discussion with the surgical service and careful review of the patient records are important. In addition, a thorough airway examination and talking with the family will aid in the complete assessment of the child.

The major complications related to surgical excision of lymphangioma include a 20% overall incidence of permanent cranial nerve injury.[6] Additional studies report a 12% incidence of permanent facial nerve paralysis in at least 1 major branch.[7] Other nerves potentially at risk for involvement during surgical excision of cystic hygroma include the accessory nerve and the recurrent laryngeal nerve. Nerve involvements also led to complications such as dysphagia, dysphonia, and difficulty with breathing.[7] Of 67 patients identified in 1 study, 88% had involvement of the oral cavity, tongue, and larynx, with 5 requiring tracheotomies.[6]

A study by Orvidas and Kasperbauer[6] indicated the recurrence rate for lymphangiomas was 39%. The high percentage of oral involvement correlated with a high degree of persistent disease.[4] The most common sites for recurrence are the tongue and hypopharynx and/or larynx. The anatomical locations of these recurrences have direct implications for anesthesia, as often the sites of recurrence are treated surgically. Multiple surgeries and frequent infections in the airway can lead to scarring and fibrosis, which can further block lymphatic drainage.[7,8] In the present case, the fibrosis of the tongue and epiglottis may have been related to a combination of the lymphangioma and to earlier surgeries. The decreased mobility seems to have contributed to the inability to displace the tongue with the laryngoscope and visualize the airway. When lymphangioma involves the tongue, it may not only harden but also enlarge the tongue.[9,10] This patient showed involvement of her tongue, inside cheek, and inner lip. Visually, there were small papillae protruding from the involved areas that were friable and bled when the oral airway was placed.

A key to caring for patients with lymphangioma is communication. The child described in this case report had a team of healthcare providers care for her since birth. There is a wealth of knowledge that can be found in direct communication in addition to the chart reviews. It is important to establish what anatomical areas are involved, the extent of the surgeries, and coexisting symptoms. The surgeon may be able to give insight as to how the lymphangioma has distorted the airway. As the child grows older, there may be a need for surgeries unrelated to the lymphangioma. While obtaining the patient's history, special emphasis should be placed on the patient's airway and the way the lymphangioma has been treated. An open line of communication between the surgeon and the anesthesia provider regarding the individual's anatomy and symptoms will increase the safety of the anesthesia provided.

Communication with parents is also vitally important. The parents know the child's current symptoms.

They may be able to answer questions relating to the patient's snoring and ability to lie flat when sleeping. Snoring may indicate a tendency for airway obstruction as the muscles of the tongue and pharynx relax during induction.[7] Symptoms of dysphagia may indicate a decreased ability to protect the airway after extubation. A complete history of recent upper respiratory infections is also necessary. It has been found that respiratory infections may exacerbate the lymphangioma by causing edema and enlargement.[7,11]

Educating the parents on their child's potential airway difficulty is important. In the present case, the child's difficult airway was discussed with the parents. The parents were encouraged to convey this information to healthcare providers before future surgeries and to obtain a Medic Alert bracelet for their child.

Advance preparation of airway equipment before surgery for a patient with lymphangioma is a necessity. Lymphangioma and previous treatments can affect the patient's airway in multiple ways. The room set-up must be individualized with a variety of oral airways and laryngoscope blades. Multiple sized oral airways should be available when anesthetizing patients with lymphangioma. This 9-year-old child required a number 9 adult oral airway because of the size of the base of the tongue. Multiple sizes of Miller blades should be available because the tongue and hardened airway structures may be difficult to move from view. Because of dysphagia and possible bleeding, suction should be functioning and immediately available; when it is used, care must be taken to avoid mucosal trauma. A tracheostomy tray should be in the operating room.

Intravenous induction versus mask induction should be discussed. Having an IV prior to mask induction would allow for increased safety if succinylcholine were needed. Cooperation of the child and support of the parents will be a factor in the induction decision-making process.

The airway examination before anesthesia needs to be individualized. The involvement of the oral cavity, oropharynx, and supraglottic structures by lymphangioma and treatments directed toward these areas may result in distortion of the anatomy. The residual lymphangioma may demonstrate mucosal friability that, when traumatized, will result in bleeding. Evaluation then relies on direct examination of the anterior oral cavity and the oropharynx. If there is evidence of involvement of the tongue base and/or supraglottic larynx, fiberoptic examination may be needed for further examination.

Conclusion

Anesthesia for the child with lymphangioma involving the oral cavity and oropharynx requires preparation and vigilance. Knowledge of the disease, previous surgeries, and the affected airway structures will help the anesthesia provider prepare for the case by having the correct equipment and the safest plan for the anesthetic. Significant airway involvement should lead to anticipation of possible airway obstruction after extubation. The patient must be fully awake, able to do a full head lift, follow commands, and swallow before extubation. Decisions regarding the preanesthetic airway plan will need to be individualized to the child. Key to the safety of the child is a thorough review of the history, communication with the surgeon, and skillful atraumatic intubation.

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