Introduction

Lymphangioma, or cystic hygroma, is a congenital malformation of the lymphatic system with an unclear embryological origin and a similar incidence rate in boys and girls. These tumours are usually localized in the head and neck. The disease has a very low incidence rate (1.2-2.8 per 1000 live births) and is usually diagnosed at birth or within the first 2 years of life. This disease usually localizes in the posterior triangle of the neck, with extensions over the parotid region and into the groin. When localized in the anterior triangle, tumours generally extend up to the mandible; extensions into the floor of the mouth and the base of the tongue are common. Cystic hygromas of the tongue are the leading cause of macroglossia in the pediatric population.

Lymphangiomas can manifest as single large cysts that can be enucleated easily, or may contain multiple cysts, which is the most common presentation. The treatment of lymphangiomas becomes urgent if the cyst enlarges rapidly due to trauma, infection, or bleeding into the cyst. Sudden enlargement can cause airway obstruction, especially if the lesion is situated in close proximity to the vocal cords, larynx, epiglottis, or tongue.

To date, several methods have been used to treat lymphangiomas, including observation, aspiration, intralesional injection of sclerosing agents (for instance, OK-432-picibanil), radium treatment, bleomycin, interferon, and steroids. There are reports of the use of OK-432 with either laser or radiofrequency ablation.

Surgery is the cornerstone of treatment of lymphangiomas; however, surgery can be challenging due to the close relationship between the cysts and important neurovascular structures. The indications for lymphangioma surgery include the mass itself, hemorrhage, dysphagia, speech disorders, dysnea, airway obstruction, infection, neuropraxia, mandibulofacial asymmetry, skeletal deviation or bony erosion, aural or ocular involvement, and aesthetic disfigurement without functional impairment. Tumour size and growth pattern are classified according to the largest diameter of the lesion in horizontal and vertical planes as assessed by pre-operative magnetic resonance imaging scans, computed tomography scans, ultrasound examinations, and histological specimens obtained during surgery: size I, 3-5 cm largest diameter; size II, greater than 5 cm in largest diameter without invasive growth pattern; and size III, greater than 5 cm with invasive growth pattern. Affected regions are categorized as neck, tongue and oral cavity, parotid and buccal area, other regions, and several regions involved concomitantly. Completeness of surgical removal is categorized as total removal, subtotal removal, partial removal, and incision and aspiration.

Herein, we present a case of difficult airway management due to protruding macroglossia in a child with lymphangioma.
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Case Report

A 5-year-old female patient presented to the Otorhinolaryngology ORL clinic suffering from a mass in her tongue and submental region (Figure 1). The lesions developed over a 30-day period. Her parents provided the patient’s history: she was born with bilateral congenital masses in her neck and she had presented to a different medical center when she was 1 year of age because of difficulty breathing. At that time, the cyst contents were aspirated twice to relieve her complaints; however, the mass did not transilluminate and it became semisolid following the second aspiration. Later, she underwent surgery to remove the mass. Bilateral incisions were made parallel to the anterior borders of the sternocleidomastoid muscles, and the cysts were enucleated. She was followed-up in the intensive care unit and was extubated on the 10th post-operative day. She did not experience any important medical problems afterwards.

When she presented to our center, physical examination revealed nasopharyngeal adenoid hypertrophy and hypertrophic pharyngeal tonsils. She had macroglossia and her tongue was protruding out of her mouth approximately 4 cm. There was an incision scar on her neck related to her operation 4 years earlier. Her laboratory test results were within normal limits.

The patient and her family were informed about the probable risks of surgical removal of the abnormal tissue, as well as anaesthetic management and post-operative care before the operation. The patient’s parents provided written informed consent.

Upon arrival in the operating room, the patient was monitored by 3-channel electrocardiography, percutaneous pulse oximetry, non-invasive blood pressure measurement, and end tidal capnography in standby. In addition, an intravenous line was inserted. Based on the clinical and radiological findings, significant difficulty in airway management was anticipated.

The patient was pre-oxygenated by facemask for 3 minutes and 1 mg midazolam was administered intravenously. Although she had a protruding tongue, the facemask fit properly and her airway was maintained. After another 3 minutes of pre-oxygenation with 100% oxygen, anaesthetic was induced with 30 mg of propofol; 10 mg of succinylcholine was administered to achieve muscle relaxation to ensure fast and gentle intubation. We used a #3 MacIntosh blade for laryngoscopy. The protruding part of the tongue was stiff; therefore, manipulating her tongue to obtain a clear view of the vocal cords was difficult. The mass and the base of the tongue were lateralized gently, and the epiglottis was seen between the hypertrophic pharyngeal tonsils. When the vocal cords were visualized, intubation was achieved with a #4.5 spiral coated Rusch orotracheal tube. Intubation was uneventful, but was highly stressful until accurate placement of the orotracheal tube was confirmed by capnography and auscultation of the chest wall with a stethoscope. Anaesthesia was maintained by 2% sevoflu-
rane in a 50% oxygen-nitrous oxide mixture. Vecuronium bromide (1 mg) was administered intravenously to maintain muscular relaxation.

The operation commenced once the patient’s airway had been secured and lasted 1.5 hours. Vital signs and oxygen saturation remained stable throughout the procedure. During completion of the operation, while the mass lesion localized anterior of the tongue was excised and the submental mass was enucleated with its capsule, we administered 0.15 mg of atropine sulfate and 0.2 mg of neostigmine to reverse muscle relaxation. The patient was extubated when she was able to hold her head and neck upright and responded to questions correctly. The patient’s oxygen saturation was above 95% after extubation and she was transferred to the intensive care unit for 24 hours of close monitoring and observation. She began to drink fluids on the second post-operative day and to eat solid food on the third post-operative day. There were no complications during follow-up. She was discharged 5 days after the operation and to date there have been no recurrences.

**Discussion**

This case concerns the anaesthetic management of a 5-year-old patient with lymphangioma. Five years is an unusual age for lymphangioma removal, these tumours are generally diagnosed and removed within the first 2 years of life. Here, we discuss the possible problems in airway protection, drug choice, and post-operative follow-up associated with surgical removal of this type of lymphangioma.

Communicating with the family and the child about the anaesthetic management during the pre-operative visit is essential. The family should be informed about the risks and problems that could occur before, during, and after surgery. During this consultation, the patient and the family can provide important information about the medical status of the patient. If the patient has a history of snoring, upper airway obstruction is a predicted outcome following the induction of anaesthesia because the muscles of the tongue and larynx relax with induction. A history of dysphagia should cause concern, because food particles can remain in the oral cavity and obstruct the airway following the induction of anaesthesia. A history of recent upper airway infection is also important because this condition can exacerbate the clinical status of the patient and can cause lymphangioma cysts to enlarge.

It is essential that the anaesthesiologist, surgeon, and nursing staff coordinate their efforts and remain in close communication. The surgeon should consult the anaesthesiologist about the radiological findings of the tumour and airway, the nature of the mass and neighboring tissues, and, as in our case, scars from previous surgeries that could affect the anatomical localization of the larynx and other upper airway structures.

In addition, the anaesthesiologist must be prepared to manage difficult airways according to American Society of Anaesthesiologist (ASA) guidelines to avoid an airway disaster. Airways of all sizes and types (oral and nasal), intubation tubes, guides, and laryngoscope blades must be prepared; suction must be immediately available and a flexible laryngoscope must be accessible. A retrograde intubation set, equipment for transtracheal jet ventilation, and tracheotomy and cricothyrotomy sets must be present in the operating room, and the ORL surgeon must be prepared for an emergency tracheotomy. In the present case, we used an adult-size airway and adult-size Miller laryngoscope blade because the patient had macroglossia.

Although awake fiber optic intubation is the gold standard for difficult intubations, we first attempted regular intubation. The equipment needed for a possible difficult intubation was available in the operating room. Propofol, which has bronchoprotective properties and a shorter half-life than sodium pentothal, was the hypnotic agent used to induce anaesthesia. Succinylcholine allows fast, repeated intubations and was therefore chosen as the muscle relaxant. Vecuronium bromide was used to maintain muscular relaxation. We administered intravenous atropine sulfate and neostigmine to reverse muscular relaxation at the end of the surgery.

In conclusion, in cases of lymphangioma, anaesthesiologists must be prepared for difficult intubations especially if is the mass is located in the neck and near the upper airway. One should be prepared for the worst possible situation and a need for tracheotomy; however, the basic procedure must be attempted first. This algorithm worked well for our patient. Another important consideration following extubation is the potential for airway obstruction due to post-operative edema. Therefore, patients should be observed in the intensive care unit and their vital signs must be monitored closely.
References


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