Received: 12.10.2008 **Accepted: 21.3.2009**

Case Report

Cystic hygroma: anesthetic considerations and review

Mohammad Reza Haji Esmaeili[*a], Seyed Soheil Ben Razavi[b]*
Hamid Reza Abbasi Harofteh[a], Seyed Mahmood Tabatabaii[a],
Habib Allah Hosseini[a], Mohammad Ali Sheikhi[a]

Abstract

Cystic hygroma (CH) in the cervical region presents as a challenge to the anesthetist. The anaesthetic difficulties are usually associated with CH because of tumor extension into the mouth, airway management, thoracic extension, hemorrhage, involvement of pretracheal region, Post operative respiratory obstruction and coexisting anomalies (Down syndromes, Turner syndromes and congenital cardiac defects). Disorders of the CH relevant to anaesthesia and intensive care medicine are discussed in this review.

KEYWORDS: Cystic Hygroma, Anesthesia, Lymphangioma, Lymphatic Vesssel.

JRMS 2009; 14(3): 191-195

Cystic hygroma, also known as cavernous lymphangioma, is a benign congenital malformation of the lymphatic system. It results from obstruction between the lymphatic and venous pathways, commonly, in the fetal neck, which leads to lymph accumulation in the jugular lymphatic sacs in the nuchal region. In the first trimester of pregnancy, the overall prevalence of cystic hygroma is about 1 in 100.[1-3] Possible etiologies implicated in the occurrence of CH are [4]:

- Altered dermal collagen composition (e.g., Down syndrome)
- Abnormal nuchal lymphogenesis (e.g., Turner syndrome)
- Hemodynamic alterations and cardiac dysfunction (e.g., heart defects)
- Abnormal endothelial cell differentiation

Treatment is surgical excision under general anesthesia either one or multistage resections. Anesthesia related problems are frequently encountered during the management of CH. These complex problems are posed by virtue of the lymphangiomas the more severe anaesthesia extension into the pharynx or thorax, hemorrhage during resection, involvement of pretracheal region, post operative respiratory obstruction and concurrent congenital anomalies (Down, Turner and Heart Defect).[5] The literature does not abound in the description of anesthetic management in extensive cervical CH. Disorders of the CH relevant to anaesthesia and intensive care medicine are discussed in this review.

Case Report

An 8 week-old, 4 kg baby girl was presented on 2008-08-09 for respiratory distress and large neck mass (Figure 1). The initial diagnosis was cystic hygroma. The CH covered the anterior and lateral neck, 28 × 13 cm² in size and extended down to the mid-thoracic and axilla region, with signs of respiratory obstruction and stridor. Examination revealed swelling on the left side of the tongue which laterally pushed the tongue upward. Transillumination

[a] Department of Anesthesiology, Yazd University of Medical Science and Health Services, Yazd, Iran.
[b] Department of Pediatric Surgery, Yazd University of Medical Science and Health Services, Yazd, Iran.
*Corresponding Author
E-mail: hajiesmaeili@ssu.ac.ir

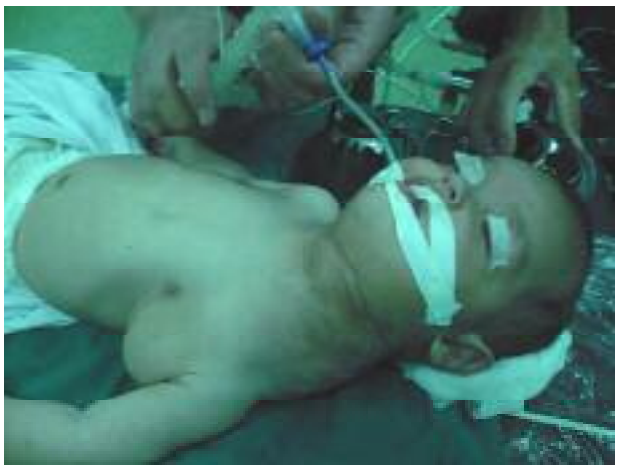

Figure 1. Photograph of patient with cystic hygroma just before surgery.

was positive. Computerized tomography (CT) of the head and neck revealed multiple fluid-filled loculi that circled the neck with mild compression and deviation of the trachea to the right. The nasogastric tube inserted earlier was aspirated. Because securing the airway under general anesthesia or a tracheostomy was considered to be difficult, the airway was secured before the induction of general anesthesia. Premedication consisted of atropine 0.12mg IV. Sedation was achieved with incremental doses of IV ketamine 1.0mg and midazolam 0.1mg to a total dose of 3.0 and 0.2mg, respectively, while oxygen was supplemented via a facemask. The larynx could not be visualized in the first attempt. Laryngoscopy was tried again and this time the glottis could be seen and a 3 mm uncuffed endotracheal tube was introduced. After ET^{CO2} confirmation of tube positioning, the ETT was taped and halothane was introduced. Ventilation was spontaneously using halothane and nitrous oxide 30% in oxygen. Blood loss was about 100 CC and replaced. The infant transferred to the pediatric intensive care unit and remained intubated for 2 days. After extubation slight deviation of the tongue was noted. After removal of dressing a sudden increasing in the size of the cyst occurs thus he was transferred to the operation room for that to aspiration of the fluid to avoid any respiratory compression. The parents were informed about possible recurrence and asked to bring the child for follow up.

Discussion

In the clinical setting lymphatic pathways can be disrupted by many different causes including congenital anomalies, infection, malignancy, radiation, surgery, and trauma. These lymphatic tumors progressively enlarged without any pain or tenderness and are often noticed in the neck. They can present as symptomatic or asymptomatic mediastinal masses. They can result in upper and lower airway compression,[6-8] as well as superior vena caval obstruction. In this article the anaesthetic implications of these conditions have been reviewed.

Preoperative Evaluation

The size and extent of the neck mass should be defined carefully in an effort to detect the potential for airway compromise and to avoid soft tissue trauma during intubation.[9] In the absence of respiratory distress, cough, tachypnea, retraction and stridor physical examination should be made for thoracic and oral extension of the tumor.[10] Inspiratory stridor suggests supraglottic obstruction, while expiratory stridor is associated with subglottic/intrathoracic obstruction.[9] All cases must have chest x-ray to exclude the presence of intrathoracic lesions.[10] Should the tumor be found in the mediastinum, further delineation with fluoroscopy,[11] angiography and CT scan [12] may aid in defining cardiopulmonary involvement and changes with respiration.[10] Anatomical and physiological differences between these patient airways and respiration was seen.[11] Concurrent disease must be evaluated.[13] Although if the patient is diagnosed at birth, the lesion is located in the anterior cervical triangle, are not associated with other birth anomalies, and generally do not required emergent surgical resection.[9]

Preparation

In Pre-operative preparation of the patient informed consent about the risks involved

should be discussed with the parent.[13] If the tumor interfered with swallowing because of extension to the mouth, the child may be malnourished or dehydrated, intravenous therapy is then required before the surgery.[14]

A full range of pediatric airways, including nasal and laryngeal mask airways should be available, as should a full range of laryngoscopes including straight and curved blade types and the McCoy laryngoscope. Expert assistance is essential. In addition to experienced operating department practitioner or nursing assistance, it is often helpful to enlist the help of another experienced anaesthetist. The facility for emergency tracheal access (e.g. cricothyrotomy kits) should be immediately available. A surgeon should standby during induction to do tracheostomy, if required.[15]

Premedication

Atropine can be given either orally or intramuscularly in a dose of 20 µg /kg to dry secretions. The use of sedative drugs to produce anxiolysis should be balanced against the risk of exacerbating airway obstruction and should be used with extreme caution.[13]

Awake Intubation

This entails direct laryngoscopy and intubation after the administration of atropine 20 µg /kg as previously described. An oxyscope, which administers oxygen during laryngoscopy, is useful. Awake intubation techniques are potentially traumatic, especially when the laryngeal structures are not visible and blind intubation is performed. There are also concerns about stress-induced physiological changes such as increases in blood pressure, heart rate, oxygen consumption and anterior fontanelle pressure, which may increase the risk of intracranial hemorrhage in premature infants.[13]

Fibre-optic intubation is performed using an ultra-thin bronchoscope over which a 2.5 mm tracheal tube can be railroaded. This technique can be performed through a laryngeal mask airway if preferred or directly via the oral or nasal routes with anaesthesia maintained via a nasopharyngeal airway. Rigid bronchoscopic equipment may also prove useful in cases where fibre-optic techniques prove difficult. The Stortz ventilating bronchoscope (Karl Stortz UK Ltd, Dundee, UK) can be used to maintain anaesthesia and oxygenation during visualization of the larynx. A bougie is threaded through the bronchoscope into the trachea and the bronchoscope removed. An appropriately sized tracheal tube can then be railroaded into the tracheal. Should these techniques fail, a surgical tracheostomy will be necessary.[13]

Partial aspiration of the cyst might facilitate intubation but would render surgery more difficult.[15]

Induction and Maintenance of Anesthesia

Induction of anaesthesia can result in the 'cannot intubate, cannot ventilate' situation or complete loss of the airway.[13] Ketamine was chosen because it preserves airway patency and causes bronchodilation.[14]

The principle behind safe induction of anaesthesia in the difficult airway is the maintenance of spontaneous ventilation. A gaseous induction using 100% oxygen with either sevoflurane or halothane is the technique of choice. The aim is to attain a plane of anaesthesia which is deep enough to allow laryngoscopy. If the airway becomes obstructed following loss of consciousness, it can be improved by turning the patient into the lateral or even semi-prone position. A nasal airway can also be useful at this early stage and is better tolerated than an oropharyngeal airway. Should the patient become apnoeic during induction, it is important to avoid assisting ventilation. Application of continuous positive airways pressure at this time will usually maintain oxygenation until spontaneous respiration resumes. It may be difficult to obtain sufficient depth of anaesthesia for laryngoscopy because of the obstructed airway.[13] Excessively high intrathoracic pressure and a high PEEP can impede the thoracic duct flow both by direct pressure on the duct and venous hypertension.[6]

Table 1. Anesthetic considerations of cystic hygroma.

1. Preoperative evaluation
• Evaluation of concurrent anomaly (Down, Turner, Heart Defect) using CXR, CT scan
• Informed consent the risks involved should be discussed with the parent
• IV fluid
2. Extension into the mouth
• Atropine can be given to dry secretions
• Induction of anaesthesia can result in the 'cannot intubate, cannot ventilate' situation
• A full range of pediatric airways should be available
• Expert assistance
• Experienced operating department practitioner
• Another experienced anaesthetist
• The facility for emergency tracheal access
• A surgeon should standby during induction
• Awake intubation
• Fibre-optic intubation
• Partial aspiration of the cyst just before intubation
• Spontaneous respiration
3. Thoracic extension.
• PEEP can impede the thoracic duct flow
4. Hemorrhage
• The wound must be drained
5. Involvement of pre-tracheal region
• Post operative respiratory obstruction
• Don't extubate
6. Observation for complication

Complication

Supraglottic edema can be prevent and treat using dexamethasone and nebulized racemic epinephrine with moist oxygen.[15]

The Management of a tracheostomy is always difficult and is worse when an emergency tracheostomy has been made. There is always considerable drainage of lymph and infection is inevitable.[13]

The wound must be drained to prevent postoperative obstruction of upper airway because of hematoma formation. Reactionary edema may develop in the first few hours post operative and necessitate tracheostomy.[1] Surgical damage to hypoglossal nerve increases the danger from the mobile tongue.[16]

Eighty percent of patients presented with life threatening complications including infection in 27%, respiratory obstruction in 17%,and intracystic hemorrhage in 15% and injuries to neighboring structures occurred in 43% (facial nerve = 6%, recurrent laryngeal nerve = 3%, internal jugular vein = 1.5%, parotid duct = 1.5%, pharynx = 1.5%). Post operative complication including wound infection in 12% and respiratory obstruction in 1.5%, edema of the tongue. Recurrence occurred in 12% within five years of surgery. It has been noted that injuries to the 9th, 10 th, 11th, 12 th and cranial nerve may occur.[16]

Acknowledgements

The authors thanks Z.H. Khan MD, professor of anesthesiology, department of anesthesiology, school of medicine, medical sciences, Tehran University, and Mohammad Reza Samiei MD, anesthesiologist, Yazd University of Medical Sciences and Health Services.

Conflict of Interests

The authors have no conflict of interests.

Authors' Contributions

MRHE, HRAH and MASh participated in preparation of the manuscript. SSBR was the surgeon of the patient. He gathered patient's data and history and carried out articles review. SMT and HAH were the Anesthesiologists of the patient and prepared the manuscript.
All authors have read and approved the content of the manuscript.

References

1. Sharma S, Aminuldin AG, Azlan W. Cystic hygroma: anaesthetic considerations and review. Singapore Med J 1994;35(5):529-31.
2. Malone FD, Ball RH, Nyberg DA, Comstock CH, Saade GR, Berkowitz RL, et al. First-trimester septated cystic hygroma: prevalence, natural history, and pediatric outcomes. Obstet Gynecol 2005;106(2):288-94.

3. Podobnik M, Singer Z, Podobnik-Sarkanji S, Bulic M. First trimester diagnosis of cystic hygromata using transvaginal ultrasound and cytogenetic evaluation. J Perinat Med 1995;23(4):283-91.
4. Nicolaides KH. First-trimester screening for chromosomal abnormalities. Seminars in Perinatology 2005;29(4):190-4.
5. Macdonald DJ. Cystic hygroma. An anaesthetic and surgical problem. Anaesthesia 1966;21(1):66-71.
6. Mallick A, Bodenham AR. Disorders of the lymph circulation: their relevance to anaesthesia and intensive care. Br J Anaesth 2003;91(2):265-72.
7. Hammer GB, Cao S, Boltz MG, Messner A. Post-transplant lymphoproliferative disease may present with severe airway obstruction. Anesthesiology 1998;89(1):263-5.
8. Hartl DM, Roger G, Denoyelle F, Nicollas R, Triglia JM, Garabedian EN. Extensive lymphangioma presenting with upper airway obstruction. Arch Otolaryngol Head Neck Surg 2000;126(11):1378-82.
9. Motoyama E, Davis P. Smith's anesthesia for infants and children. 7th ed. Philadelphia: Mosby; 2006. p. 515.
10. Maze A, Block E. Stridor in pediatric patients. Anesthesiology 1979;50(2):132-45.
11. Perkes EA, Haller JO, Kassner EG, Wolf E, Velcek FT. Mediastinal cystic hygromas in infants. Two cases with no extension into the neck. Clin Paediatr (Phila) 1979;18(3):168-70.
12. Leipzig B, Rubuzzi DD. Recurrent massive cystic lymphangioma. Otolaryngology 1978;86(5):758-60.
13. Cardwell M, Walker RWM. Management of the difficult pediatric airway. British Journal of Anaesthesia: Continuing Education in Anaesthesia, Critical Care & Pain 2003;3(6):167-70.
14. White PF, Way WL, Trevor AJ. Ketamine-its pharmacology and therapeutic uses. Anesthesiology 1982;56(2):119-36.
15. Jaffe RA, Samuels SI. Anesthesiologist's manual of surgical procedures. 2nd ed. Philadelphia: Lippincott Williams and Wilkins;1999. p. 952-4.
16. Ameh EA, Nmadu PT. Cervical cystic hygroma: pre-, intra-, and post-operative morbidity and mortality in Zaria, Nigeria. Pediatr Surg Int 2001;17(5-6):342-3.