Cystic hygroma in the neck causing airway compromise: Case report and review of literature

Cheuk-Chung AU,1 Shuk-Kuen CHAU,1 Hau-Yee CHAN,1 Nai-Shun TSOI1
1Department of Paediatrics and Adolescent Medicine; 2Department of Surgery, Queen Mary Hospital, Hong Kong

Abstract

We report a case of cystic hygroma in a 7 month old boy causing severe airway compromise, and discuss the acute management of airway obstruction and the surgical management of cystic hygroma.

Keywords: Airway management, Cystic hygroma, Lymphangioma, Sclerotherapy

Case

A 7 month old baby boy was brought to the Accident & Emergency department (AED) because of marked noisy breathing and shortness of breath. There were preceding symptoms of coryza and fever for a few days. He was born full-term with uneventful postnatal course in China. No abnormality was told for the antenatal scan. Mild snoring during sleep was noted since baby was about 4 months old but no evaluation was done at that time.

Assessment at AED revealed significant stridor and marked respiratory distress with tachypnea and use of accessory muscle. There was noticeable swelling over the left neck (Figure 1). His oxygen saturation measured in room air was only 70% hence he was given 6L/min oxygen via face mask. In view of the severe airway compromise, emergency team was activated including paediatrician, otorhinolaryngologist and anaesthetist. Baby was escorted to the operating theatre and the airway was secured by endotracheal intubation with 3 mm microcuffed tube under gas induction. Direct laryngoscopy revealed smooth soft cystic swelling at the oropharynx from the level of uvula down to the arytenoid and the lesion demonstrated transillumination. Ultrasonography showed multiple septation over the left neck with no significant colour flow which was suggestive of lymphatic malformation.

Emergent ultrasound-guided aspiration of the cystic lesion by paediatric surgeon yielded 57 ml clear light brownish fluid, it was then followed by injection of doxycycline into the lesion as sclerotherapy. Computed tomography of the neck with contrast showed multi-septated mildly rim-enhancing hypodense collection measuring 7.5 cm x 3.6 cm x 5.4 cm in the retropharyngeal region which extended to the nasopharynx superiorly and down to the level of the T2 vertebra inferiorly. The airway was deviated to the right and was most narrowed at the level of the thyroid isthmus. He remained intubated with mechanical ventilation in view of his severe airway compromise. A second operation was performed by paediatric surgeon after two weeks, in which 100 ml straw coloured fluid was aspirated from the lesion followed again by doxycycline as sclerotherapy. Clinically the neck swelling decreased and resulted in air leak around the endotracheal tube, and he could be extubated. However, his respiratory symptoms recurred quickly within a week and re-assessment computed tomography scan showed that despite the lesion within the retropharyngeal region had mild interval reduction in size, the airway was still partially obliterated between the nasopharynx and the oropharynx. He was then put on continuous positive airway pressure (CPAP) support with a pressure of 8 cmH20. He received two more sessions of sclerotherapy with doxycycline and another session of sclerotherapy with bleomycin. Magnetic resonance imaging performed at 10 months old, i.e. 3 months post initial presentation, revealed interval decrease in size of the left retropharyngeal and parapharyngeal lesion compared with previous imaging (Figure 2). His clinical symptoms improved but he still required CPAP support during sleep for his obstructive sleep apnea symptoms.

*Author to whom correspondence should be addressed.
Email: aucc@ymail.com
Discussion

Cystic hygroma is a form of lymphangioma and is a congenital malformation in the lymphatic system. It is now classified according to the International Society for the Study of Vascular Anomalies as "common (cystic) lymphatic malformations" under "simple vascular malformations". It could be divided into macrocystic, microcystic, or mixed cystic lymphatic malformations, where the cysts measures more than or less than 2 cm³ respectively. It is thought to originate from the failure of lymphatics to connect with the venous system, and the sequestration of lymphatic tissue in lymphatic sacs during embryological development. It may occur in different parts of the body. The head and neck area is the commonest location in up to 80% of cases. Other areas may include the axilla, mediastinum, retroperitoneum, and groin. Most cases present within the first two years of life.1,2 It presents usually as an asymptomatic mass, but it could also present with complications from the mass effect such as acute or chronic respiratory compromise and feeding problem with subsequent failure to thrive. Large cystic hygroma may extend from the head and neck into the mediastinum and compress onto the trachea.3 Inflammation in the cystic hygroma after an upper respiratory tract infection or abscess formation, or spontaneous or traumatic haemorrhage into the cystic hygroma may result in rapid increase in size.3

Figure 1. Lateral neck radiograph showing retropharyngeal swelling with anterior displacement of the airway.

Figure 2. (a) Computed tomography. Large cystic hygroma in the neck displacing the airway to the right, endotracheal tube in-situ to protect the airway. (b) Magnetic resonance imaging. Reduction in size of cystic hygroma after sclerotherapy.
Acute airway obstruction from a rapidly enlarging cystic hygroma is a serious complication that necessitates emergent airway management. A team of experienced paediatric intensivist, anaesthesiologist, and otorhinolaryngologist should be assembled to stabilize the airway under controlled environment in the operating theatre or intensive care unit. Before a definitive airway is established, the patient should be minimally handled, accompanied by the care-giver, and be allowed to stay in a comfortable position of his preference. A difficult intubation due to the airway obstruction and anatomical distortion should always be anticipated. It is prudent to maintain spontaneous respiration with careful choice of anaesthetic agents and avoid neuromuscular blockers. Alternative equipment should be available in the case of difficult intubation; this may include an optical or light stylet, a bougie, a video laryngoscope, or a laryngeal mask airway. Rigid bronchoscopy, emergency cannula cricothyrotomy with transtracheal jet ventilation, or surgical cricothyroidotomy should also be available as the rescue. Tracheotomy may be required to relieve the respiratory obstruction.

Sclerotherapy and surgery are the two major treatment modalities for cystic hygroma. Sclerotherapy works by inducing inflammation which results in resolution of the lesion. The cystic hygroma is aspirated and the sclerotherapy agent is then injected. Common adverse reactions include fever, and local pain, erythema, or swelling in the first few days; the swelling could also result in airway obstruction. Repeated injections at intervals, or combination with surgery, may be necessary. Most studies used the sclerotherapy agent OK-432, doxycycline, and bleomycin. OK-432 is a derivative from Streptococcus pyogenes and a potent immunostimulant. It had a success rate of 87% to induce resolution of greater than 50% size of a macrocystic lesion. However, it is not readily available for treatment in our locality. Doxycycline is a tetracycline antibiotic, it could also induce inflammatory reaction in the endothelial line cavity of the lesion. Study found similar effectiveness in both OK-432 and doxycycline for treatment of macrocystic lesion, although doxycycline required fewer repeated injections and produced less intense inflammatory reaction. Doxycycline had the potential risk of tooth discoloration in the young children, for which the parents should be warned. Bleomycin is a chemotherapy agent with similar effectiveness to OK-432 and doxycycline. The main serious side effect of bleomycin is pulmonary fibrosis and the total dose of bleomycin should therefore be limited.

Surgery was considered a treatment of choice for cystic hygroma. However, complete removal was often not possible with large lesion near the major vessels and aerodigestive tract, and the recurrence rate would be high. Nerve paralysis, such as facial nerve palsy or Horner’s syndrome, recurrent airway obstruction, and infection and scarring are important complications to consider. Comparison between surgery and sclerotherapy showed no clear superior treatment outcome.

Conclusion

Cystic hygroma in the neck may cause severe life-threatening airway compromise. Prompt recognition and expectant advanced difficult airway management to protect the airway in this scenario is the priority. Treatment options for the lesion with different sclerotherapy agents and surgery could be considered.

References