CASE REPORT

Dilemma in management of cervico-facial cystic hygroma

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SUMMARY
Cervico facial cystic hygroma and tongue lymphangioma is rare representative of spectrum of lymphatic malformations. Conservative management with sclerosants alone has proven to be successful. However, sudden enlargement of these cervico facial lymphangiomas leads to catastrophic airway obstruction leading to debility in feeding and speech. Therefore, surgery is indicated in such case to prevent such a catastrophic problem. We report here the case of a 3-year-old boy with cervico facial hygroma involving the tongue. We successfully treated him with a combination of surgery and OK432 injection.

INTRODUCTION
Cervico facial cystic hygroma and lymphangioma is representative of a spectrum of lymphatic malformation. The location of lymphangioma is an important factor in determining the clinical presentation, complications and outcome of surgery. Rapid growth and enlargement of cystic hygroma in cervico facial region due to trauma and infection increases mortality and morbidity of the patients. The condition rarely undergoes a complete regression. These are common benign developmental tumour in the paediatric population that pose special challenges in managing among the otolaryngologist-head and neck surgeons.

CASE REPORT
A 3-year-old boy presented with submandibular neck swelling since birth. The swelling increased in size (8x5x3cm) and this was associated with prominent protrusion of tongue since the age of one. The tongue protrusion became worse until at a stage, the tongue was three times bigger than the normal size and 2/3 of the tongue was outside the mouth. He was unable to close his mouth and faced difficulty in eating and speaking (Fig. 1). There was no respiratory distress. Magnetic Resonance Imaging of the neck demonstrated multicystic mass measuring 2.2cm x 6.3cm x 3.9cm in the neck with hemorrhagic cystic lesion measuring 2.9cm x 1.8cm x 3.5cm extending into the base of tongue causing airway obstruction. The OK-432 is the latest sclerosant agent that produced by the low-virulence Su strain of group A Streptococcus pyogenes that is treated with penicillin G potassium. Study by Ogita et al., reported that 67% of 46 cases treated with only OK432 showed total or marked shrinkage of lymphangioma. The main presentation of cervicofacial lymphangioma is a mass. It is usually not detected at birth due to the small size and only noticeable later in life due to upper airway infection and trauma. Airway obstruction, feeding and speech difficulty are common in lymphangioma involving oral cavity, pharynx and/or larynx.

Differential diagnoses are brachial cleft cyst, dermoid cyst, hemangioma, thyroglossal cyst, laryngocele, thyromass and lipoma. Most authors recommend magnetic resonance imaging (MRI scan) for the diagnosis and to plan surgery.

Management of lymphangioma remains controversial. The ideal treatment is conservative management with sclerosant injection. Other non-surgical modality includes aspiration and radiation. Aspiration can be temporary measure to reduce pressure effect on respiratory and feeding passage. Irradiation or radon seed implantations have been used but limited due poor response rate and high side effects. Sclerosant injection agents include steroids, alcohol, bleomycin sulphate, tetracycline, and OK432.

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Another study observed an excellent result with only OK432 injection whereby 90% had complete regression within 1-2 months. They hypothesized that sclerosant damages the endothelial lining and cause obliteration of the lymphangioma cavity, hence prevent accumulation of fluid within the lesion. The initial local immune response of OK432 will result in fever, pain, rapid temporary increase in size of cystic hygroma.

Surgery is indicated when there is increase in size, frequent infections, debilitating functions or/and life-threatening symptoms. As in the present case, surgery was chosen due to child having feeding and speech difficulties. A complete excision of cystic hygroma is almost impossible as in this case without sacrificing the important neurovascular structures that are closely related. Even though it is sensible to save these structures, leaving behind residual cyst will increase the recurrence rate.

Location of lymphangioma is predominant factor in surgical outcome. Literature reviews conclude in increased post-surgery recurrence rate, morbidity and complications in surgery of suppharyoid lesion. Also, those lesions with mucosal involvement like floor of the mouth and tongue resulted in poor surgical outcome.

Also, airway and swallowing problems may arise or persist after surgery due mucosal oedema, damage of neural innervation to the tongue or pharynx and enlargement of internal lymphangiomas. For example, excision of bilateral submandibular hygroma or tongue reduction surgery has higher risk in developing post-operative lingual oedema due to interruption of lingual lymphatic. Therefore, in our case, tracheostomy was undertaken by anticipating post-surgery airway obstruction.

CONCLUSION
It is crucial for surgeons to understand the nature this disease and be able to weigh the ideal management based on their surgical experience and limitations present. A combination of surgery with sclerosant agent like OK432 has excellent outcome with minimal complications.

PATIENT CONSENT
Permission for patient photographs obtained from mother (legal guardian).

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REFERENCES