CASE REPORT

Dilemma in management of cervico-facial cystic hygroma

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SUMMARY

Cervico facial cystic hygroma and tongue lymphagioma 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 cervicofacial region due to trauma and infection increases mortality and morbidity of the patients.[1] 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 protrusion of the tongue (Fig. 2). A diagnosis of cystic hygroma with tongue lymphangioma was made. He underwent excision of the swelling, partial glossectomy and tracheostomy. Intra-operatively multiple cystic mass involving bilateral submandibular region, floor of mouth and tongue was noted. No clear plane was seen. The neck lesion was excised through a submandibular approach till the floor of mouth. The surgery was uneventful with removal of more than 90% of the swelling. This was followed by an ultrasound guided OK432 injection in a month post operatively. After three years, patient was able to close his mouth fully and tolerate food and able to speak.

DISCUSSION

Cervico-facial lymphangioma is a congenital benign lesion. It arises from localised lymphatic stasis due to sequestration of lymphatic tissue derived from portions of primitive sacs during embryology life.[2] A widely used classification by Landing and Faber categorises these lesions into capillary lymphangioma/lymphangioma simplex, cavernous lymphangioma and cystic lymphangioma/cystic hygroma based on size and vascular spaces.[1] In some patients, the lesions may appear together depending on the severity.

The main presentation of cervicofacial lymphagioma 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.[2] 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, thymic cyst, thyroid mass and lipoma.[2] Most authors recommend magnetic resonance imaging (MRI scan) for the diagnosis and to plan surgery.[2]

Management of lymphagioma remains controversial. The ideal treatment is conservative management with sclerosant injection. Other non-surgical modality includes aspiration and radiation.[2] Aspiration can be temporary measure to reduce pressure effect on respiratory and feeding passage.[3] Irradiation or radon seed implantations have been used but limited due poor response rate and high side effects.[1] Sclerosant injection agents include steroids, alcohol, bleomycin sulphate, tetracycline, and OK432.[2]

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.[4] Study by Ogita et al., reported that 67% of 46 cases treated with only OK432 showed total or marked shrinkage of lymphangioma.[4]

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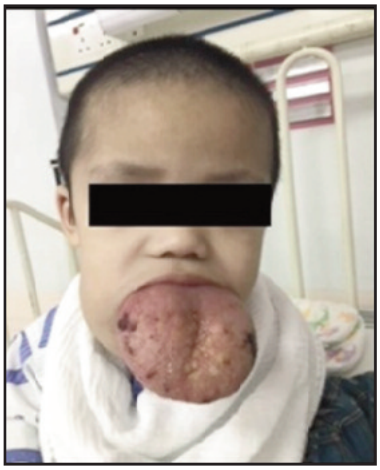

Fig. 1: Prior to treatment.

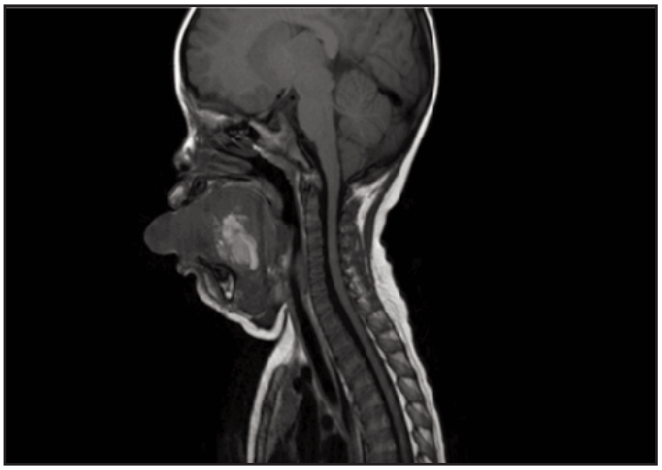

Fig. 2: weighted MRI head and neck (sagittal) Shows haemorrhagic cystic lesion extending into base of tongue causing protrusion of tongue.

Another study observed an excellent result with only OK432 injection whereby 90% had complete regression within 1-2 months.[4] They hypothesized that sclerosant damages the endothelial lining and cause obliteration of the lymphangioma cavity, hence prevent accumulation of fluid within the lesion.[4] The initial local immune response of OK432 will result in fever, pain, rapid temporary increase in size of cystic hygroma.[2]

Surgery is indicated when there is increase in size, frequent infections, debilitating functions or/and life-threatening symptoms.[1,2] 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.[5] Even though it is sensible to save these structures, leaving behind residual cyst will increase the recurrence rate.[5]

Location of lymphangioma is predominant factor in surgical outcome.[1,2] Literature reviews conclude in increased post-surgery recurrence rate, morbidity and complications in surgery of suprahyoid lesion.[1,2] Also, those lesions with mucosal involvement like floor of the mouth and tongue resulted in poor surgical outcome.[2]

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.[2] 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.[1] 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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