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Lymphovenous malformation of tongue: airway challenge

R. D. Chhabria, N. M. Dave, M. Garasia

Department of Anaesthesiology Seth GS Medical College and KEM Hospital, Mumbai, India

Corresponding author: R. D. Chhabria, Department of Anaesthesiology Seth GS Medical College and KEM Hospital, Mumbai, India Email: drach_18@yahoo.com

Key points

Lymphovenous malformations involving the aerodigestive tract pose a significant challenge. Anaesthesiologist needs to be aware of the treatment modalities, their complications, and anticipate and plan accordingly to manage them. Intubation and extubation of difficult airway should be done in a well equipped facility. Supportive treatment in form of short acting steroids, anti-inflammatory agents, antibiotics, play a vital role in managing inflammatory reaction post use of sclerotherapy agents. An interdisciplinary approach involving different specialities including paediatricians, interventional radiologists, anaesthesiologists, intensivists, ENT surgeons is crucial to management of such conditions.

Abstract

Lymphovenous malformations are sequestrations of developing lymphatic system, located in head and neck in most cases. Involvement of the tongue poses an airway challenge. We present a case of lymphovenous malformation of tongue in a ten year old child with speech and breathing difficulties. This report illustrates the specific difficulties encountered and its perioperative management with meticulous planning using multidisciplinary approach.

Keywords: Lymphovenous malformation of tongue, difficult airway, sclerotherapy

Introduction

Lymphovenous malformations are birth defects, formed by sequestration of developing lymphatic system. 50 % are recognized at birth and 90 % by the age of two years.[1] 60 % are located in head and neck.[1] Malformations of the airway are a challenge to deal with. We present a case of a ten year old male child with lymphovenous malformation of the tongue causing airway compromise. This report illustrates the difficulties that may be encountered while handling such malformations and the involvement of anaesthesiologist in their management.

Case report

A ten year old male child was referred to our hospital for evaluation of a gradually increasing swelling in the floor of mouth and tongue, present since birth. There was history of sudden increase in size of the swelling since few months causing tongue protusion leading to difficulty in closing mouth, feeding, speech and breathing. On examination a large, non-tender, compressible swelling in bilateral submandibular and submental region, involving the tongue was noted (Figure 1). CT scan revealed large hypodense lesion involving bilateral submandibular regions, floor of mouth and tongue with multiple large septae. MRI showed extensive slow lymphovenous malformation with epicentre in floor of mouth (Figure 2). High frequency sonography of neck showed partially compressible multiple septated lesion with anechoic cystic component in floor of mouth 5.5cm×5.8cm×6.8cm. Lesion had few interspersed small calibre vessels communicating with external carotid artery branches without significant increase in vascularity.

The lesion closely abutted bilateral submandibular gland without invasion, extending into genioglossus muscle and base of tongue. Vocal cords, thyroid gland, parotid gland and sublingual gland were not involved. Chest X-ray, ECG, 2DECHO were normal. The child was on tab.propanolol 10 mg BD.

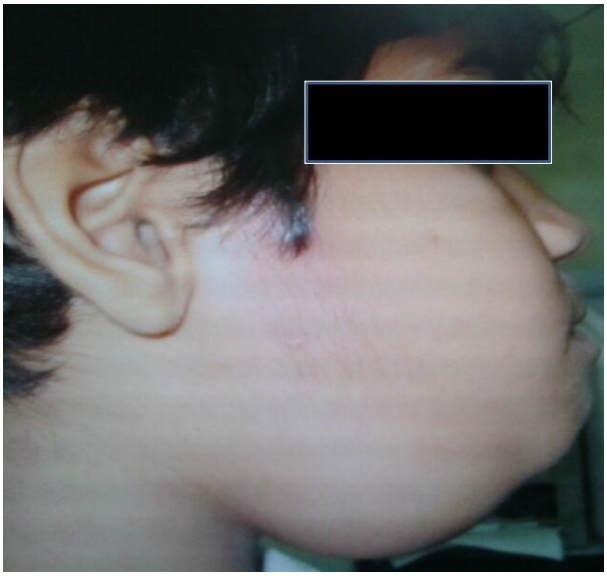

Fig. 1. Lateral picture showing extent of lesion

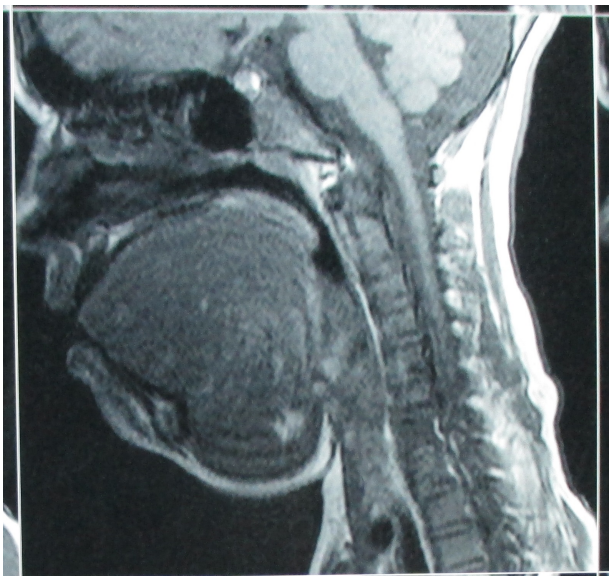

Fig. 2. MRI showing lymphovenous malformation

Child was posted for cerebral angiography and further management depending on extent of the lesion. Anticipating difficult airway, awake fibreoptic intubation was planned. Glycopyrolate 0.2 mg IM was administered. Topical analgesia of upper airway was achieved with lignocaine 2% nebulisation and viscous 4% lignocaine gargles. Child was intubated nasally with the flexible fibreoptic bronchoscope with 5.5 mm endotracheal tube. After intubation the child was anaesthetised and muscle relaxant was given. A direct laryngoscopy done after intubation revealed Cormack and Lehane grade I airway. Cerebral angiography revealed bilateral internal carotid artery, external carotid artery and vertebral artery as normal and no evidence of abnormal vascular blush or hypertrophied feeding artery. The interventional radiologist decided to use sclerotherapy for the lesion and injected 3cc of sodium tetradecylsulphate (STD), non-ionic contrast into the lesion. Post procedure anticipating inflammatory reaction, the child was kept intubated and shifted to paediatric intensive care.

Post procedure, the lesion was severely inflamed protruding outside the oral cavity. The inflammatory reaction was conservatively managed. Antibiotics, non-steroidal anti-inflammatory drugs plus short acting steroids formed the main stay of therapy. Supportive therapy in form of regular ETT suctioning, nebulisation and chest physiotherapy was done in the ICU. On the third post procedure day, after the inflammation subsided, child was extubated after shifting in the operation theatre. Anticipating difficult extubation, extubation was done over an airway exchanger and with tracheostomy standby with ENT surgeons inside the theatre. The child was given tab.propanolol 10 mg OD for next two months and posted for second sclerotherapy session.

For the next session the lesion had decreased in size and the child was intubated nasally under direct laryngoscopy. 35cc of STD+15U Bleomycin was injected into the lesion. With previous experience, the child was kept intubated. The lesion inflamed more than the first time and the child was conservatively managed and extubated on 4th day after the procedure. The child received one more session of sclerotherapy with 20U Bleomycin which resolved the swelling completely.

Discussion and conclusion

Cystic hygroma also called as cavernous haemangioma, Lymphovenous malformations (LM) may be diffuse or localized and cause symptoms depending on its location. LMs of head and neck can cause significant airway compromise requiring urgent intervention with intubation or tracheostomy, if symptoms like severe respiratory distress or stridor occur.[2] Depending on the size, LMs are classified as Macrocystic or Microcystic. Macrocystic lesions are soft, compressible, transilluminate, found in neck and have good prognosis. Microcystic lesions are usually present at birth causing distortion of cervicofacial soft tissues, even maxillofacial bones.[1, 2] Treatment modalities mainly include sclerotherapy or surgery.[2, 3]

LM of the tongue leads to macroglossia, which causes cosmetic deformity, speech disturbances, difficulty in feeding, airway obstruction, orthodontic abnormalities-mandibular prognathism and malocclusion. It may get complicated with infection or haemorrhage.[1] Direct malformation involvement of the upper aerodigestive tract can cause significant functional compromise that is difficult to treat.[4]Lymphatic malformation treatment improvements have been made through radiographic characterization, percutaneous sclerotherapy with STD and Bleomycin are considered as the most effective and safe treatment modality for low flow vascular malformations involving the head and neck.[3,4] Saraf S. et al in analysing the role of sodium tetradecyl sulphate in venous malformations found that all patients experienced pain and swelling to a variable degree which lasted typically for one to two weeks.[5] Management of such lesions are complex and require interdisciplinary approach involving different specialities like paediatricians, intervention radiologists, anaesthesiologists, intensivists, ENT surgeons, orthodontists, psychologists and speech therapists.

Anaesthesiologists could be involved intraoperatively or post operatively, while handling such cases. A number issues need to be tackled in this case including difficulty airway due to macroglossia, difficult ventilation so awake fibreoptic intubation with tracheostomy standby was planned. Difficult airway options available in children like Bullard Laryngoscope, Airtraq Optical Laryngoscope, Glidescope, Storz Video Laryngoscope, Truview require some mouth opening and are of limited utility in oral lesions.[6] Fibreoptic bronchoscopy is considered to be the gold standard.[6] Postoperative conservative ICU management maintaining patency of ETT avoiding tracheostomy was important. Anticipating difficult extubation, it was planned inside OT and done over an airway exchanger with tracheostomy standby. The anaesthesiologist needs to be aware of the treatment modalities and their complications.

With thorough history, investigations, anticipating difficulty, planning and preparing for difficult airway and post operative management and with interdisciplinary approach, LMs of the airway can be safely and successfully managed.

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