Cystic Hygroma with Extensive Tongue Involvement

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Abstract Cystic Hygroma is a congenital lymphangiomatous tumour. It commonly involves the neck but is rarely associated with lymphangioma of the tongue. Presenting here a rare case having extensive tongue involvement giving a terrifying look and severe anaemia but without respiratory distress. Herewith submitting a case report with a brief review of literature and various problems faced while managing the same.

Keywords Cystic hygroma · Macroglossia · Lymphangioma

Introduction

Cystic hygroma is a congenital lymphangiomatous tumour originating from sequestered lymphatics from the primitive jugular sac [1]. They commonly present very early on in life as a neck swelling involving posterior triangle of neck. Small lymphangiomas usually present only as a cosmetic deformity but larger swellings may compromise airway or interfere with feeding and swallowing [1, 2]. Anterior triangle cystic hygromas involving the suprahyoid compartment may infiltrate into tongue causing lymphangiomatous cysts in the tongue.

Case Report

A 10 year old male child attended the ENT OPD of GMC Nagpur with complaints of a neck swelling since birth and protrusion of tongue since past 3 days. The patient was unable to close the mouth or retract the tongue inside the mouth (Fig. 1).

On clinical examination the neck swelling was large, about 20 × 8 cm in size, extending from one angle of mandible to another and from chin down to the thyroid cartilage. Local temperature at the site of swelling was raised and slight tenderness on palpation could be elicited. The swelling was neither compressible nor reducible. Fluctuation could be elicited over the swelling. The tongue was protruded outside the oral cavity by about 5 cm. It was grossly thickened and indurated giving a terrifying look. Bite marks due to upper and lower incisors were present over the tongue with minimum oozing of blood from the site. The patient surprisingly did not have respiratory distress but coincidentally he was severely pale.

The child was immediately investigated and Haemoglobin was found to be only 2 g%. Plain radiograph of the neck, lateral view, showed a soft tissue swelling in the neck with adequate airway (Fig. 2). Radiograph of neck with chest showed that the swelling was essentially limited to the neck without any mediastinal extension (Fig. 3). USG of the neck revealed a multiseptate loculated cystic lesion with intralobular haemorrhage within few of the loculi. The tongue was grossly oedematous but the vascularity of the tongue was well maintained. However no comment was made about the continuity of the neck and tongue swelling. A CT scan could not be done as the patient could not afford it.

On the basis of above clinical findings a provisional diagnosis of

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Fig. 1 Picture showing neck swelling with macroglossia

Fig. 3 Radiograph of neck lateral view showing adequacy of airway

Fig. 2 Picture showing discoloration of tongue

- (1) Cystic Hygroma with tongue lymphangioma with severe anaemia or
- (2) Cystic Hygroma with angioneurotic oedema of the tongue with severe anaemia, as such a huge macroglossia with a history of short duration of history had not been encountered in past 19 years of practice.

In view of this diagnosis steroids along with blood transfusion were administered.

Over the next 24 h, there was no sign of reduction in the size of the tongue swelling, instead discoloration of the tongue had began to be obvious (Fig. 4). Hence decision was taken to operate upon the child as an acute surgical emergency though the Hb levels were very low.

Tracheostomy was done preoperatively as oral intubation was impossible due to the macroglossia and restricted mouth opening. Horizontal neck incision was taken in the skin crease. A multiloculated cystic lesion was dissected out. Part of the lesion was left behind in order not to

Fig. 4 Radiograph of neck anterior view showing swelling limited to neck without mediastinal extension

compromise the integrity of the floor of the mouth and as no definite surgical plane for excision could be found. Besides due to low Hb levels lengthy surgery had to be avoided. An anterior partial reduction glossoplasty was done for the macroglossia (Fig. 5) and the devitalised portion of the tongue was removed with it. Intra operative and post operative blood transfusions (total 6) were administered and haemoglobin levels restored to 10 g%. Post operatively patient was managed with intravenous antibiotics, Ryle's tube feeding and tracheostomy tube care. The post operative histopathological diagnosis was of

Fig. 5 Post operative photograph of patient

“cavernous lymphangioma (microcystic variant) with eosinophilic debris and RBCs within the cysts”.

Post operatively patient required intensive care. A decannulation of the tracheostomy was attempted but patient did not tolerate it and required ventilatory support. Patient's parents were unwilling for any further intervention at present, hence further surgery has been temporarily deferred.

Discussion

Lymphangiomas are benign hamartomatous tumors. 75% of these occur in the head and neck region [1, 3].

65% cases of cystic hygroma present within a year of birth and 90% are diagnosed within 2 years of life [1]. Rare cases have been reported where these lesions have been a cause for obstructed labour and cases can be diagnosed ultrasonographically as early as in the 2nd trimester [2].

Lesions commonly involve the posterior triangle of the neck. Anteriorly placed cystic hygroma with intraoral extension with tongue involvement is an uncommon clinical condition. Huge neck swellings present with airway obstruction and stridor requiring emergency aspiration and even tracheostomy is needed in nearly 50% cases [4]. In this case, as the tongue was protruding outside the oral cavity, there was no tongue fall and no stridor. However tracheostomy had to be done preoperatively as intubation was not possible.

The low haemoglobin levels were also a problem. However in view of the impending devitalisation of the tongue emergency surgery had to be performed.

The macroglossia could be attributed to a variety of reasons. It could be because of lymphangiomatous cysts in

the tongue, which are known to affect the anterior 2/3rd [3, 5]. It is also postulated that local infection, even simple URI, or intralesional haemorrhage can give rise to sudden increase in the size of a pre-existing lymphangiomas [2, 6] which can explain the short history of tongue protrusion. Also the neck mass itself could mechanically displace the tongue out of the oral cavity causing its protrusion.

For the macroglossia, anterior partial reduction glosso-plasty was performed [4, 7]. This is a documented modality for management of lymphangiomas involving the tongue. Also the extent of the neck excision was a dilemma. Here it is important to note that this is essentially a benign condition and normal anatomical structures should not be compromised in pursuit of complete excision and in such cases subtotal surgery is acceptable with revision surgery at a later setting [1]. Apart from surgery, other available options are LASER vaporisation using CO₂ or Nd-Yag LASER, sclerotherapy or cryosurgery [3, 8]. Various sclerosants have been tried ranging from anti-cancer drugs like intravenous cyclophosphamide or bleomycin [2, 3], to newer agents like fibrin glue [2], or picibanil (OK-432) [2, 3] a low virulent *Su* strain of type III group A *Streptococcus pyogenes*.

Conclusion

This is a unique case, in the sense that there was no respiratory distress in spite of the macroglossia. This was so because the tongue had been pushed forward and held in position by its sheer size and by being fixed between the teeth. In addition to this the Hb was only 2 g% and devitalisation of the tongue started on the very next day of admission. This put us in a dual mind as to whether to operate immediately or wait for improving Hb and procuring further radiological investigations like CT scan. In either case there was certainly the potential danger of further ischemia of tongue. At last we decided to go for surgery and we succeeded partially, as part of the lesion at the floor of the mouth had to be left behind to prevent collapse of the floor of oral cavity and also as the general condition of the patient did not permit an extensive surgery and so patient was advised further operative work in a leisurely planned manner. But unfortunately, the patient was not willing for revision surgery against medical advice.

Such a lesion may be encountered in clinical practice which puts the treating surgeon in a difficulty in deciding line of management as described here, hence presenting this case.

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