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Case Report

CASE REPORT

Cystic Hygroma with Difficult Airway in an Adult

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ABSTRACT

Cystic hygroma (CH) is an uncommon benign lesion usually occurring in the cervicofacial region and was first described by Wernher in 1943. It is commonly seen in neonates and in early infancy. CH is rare in adults with less than 150 adult cases with only few with difficult intubation have been reported in literature and 2-6% die of airway compromise. We report a rare case of CH in a 61-year-female occupying whole of oral cavity and extending to the neck, with feeding and breathing difficulty. This was classified as de Serres stage 5 and Mallampati grade IV. This case highlights that cystic hygroma be considered in the differential diagnosis of cystic neck masses in adults and considering the issue of difficult intubation, identification and appropriate plan for airway management is key to successful outcome. In this case surgical airway was on the cards, intubation by a skilled expert anesthetist using mackintosh laryngoscope made complete surgical removal through a trans cervical approach possible.

Key Words: Cystic Hygroma, Cystic neck masses, Difficult intubation.

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Article info.
Received: September 12, 2017
Accepted: September 23, 2017

Cite this case Report: *Saqulain G, Ahmed J, Shahan Z, Nawaz N. Cystic Hygroma with Difficult Airway in an Adult. JIMDC. 2017; 6(3): 197-201.*

Funding Source: *Nil*
Conflict of Interest: *Nil*

Introduction

Cystic hygroma (CH), first described by Wernher in 1843 who coined the term, is synonymous with cystic lymphangioma. CH can affect any anatomic site in the body, majority of them occur in the Head and Neck.[1-3] Incidence is 1.2 to 2.8 per 100,000 infants.[4,5] Presentation in adulthood is rare.[6,7] Two to six percent of cystic hygroma cases usually die due to airway compromise. Patients usually present with a painless, soft, fluctuant, trans illuminant, and enlarging neck mass. A careful history, clinical examination and imaging is essential for diagnosis. MRI is the consensus study of choice however; CT scanning is faster and more readily available. Complete surgical excision is the treatment of choice, but is possible in around forty percent of cases.

We, present an unusual rare case of CH in a 61-year-old female with a difficult airway. It was a large de Serres grade 5 cystic hygroma with Mallapatti grade IV. Orotracheal intubation using a mackintosh laryngoscope by an expert anaesthetist helped avoid a surgical airway and the mass was removed successfully using trans cervical approach. Literature review reveals only a few cases of CH's with difficult intubation in adults.

The objective of this case report is to highlight that CH needs to be considered in the differential diagnosis of cystic neck masses in adults as well, and considering the fact that CH can involve the airway and result in difficult intubation, identification and plan for airway management is key to the successful outcome.

Case Report

A 61-year-old female presented to the Otolaryngology outpatient with a 40 years' history of painless slowly growing swelling neck and floor of mouth which initially started as a small submental swelling and grew very slowly over 40 years and finally presented with 6 months history of dysphagia and breathing difficulty. Patient was a naswar addict. On clinical examination, there was a huge, cystic swelling 10 x 9 cm filling the oral cavity arising from

the floor, with the tongue pushed upwards and backwards into the pharynx (figure Ia). Other intraoral structures were normal. A similar around 10 x 9 cm cystic, trans-illuminant, non-pulsatile mass was occupying the submental and submandibular areas with normal skin (figure Ib).

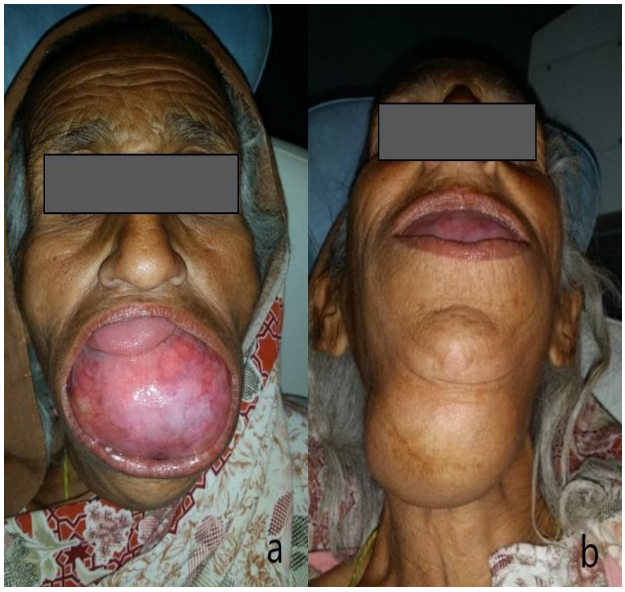

Figure I: Preoperative photograph showing a) Large CH occupying the oral cavity with wide mouth opening and b) Neck mass occupying the submental and bilateral submandibular area.

The margins of swelling were ill-defined with reduced mobility. There was no other swelling/ mass/ palpable lymph node in neck. Neurological examination was unremarkable. Computed Tomography (CT) scan with contrast (figure 2) showed a large minimally complex predominantly cystic lesion measuring 74 mm AP x 69 mm transverse with 95 mm craniocaudal length, centered in sublingual location surrounded by 6 mm thick soft tissue wall, filling the oral cavity with significant extension into the neck. Tongue pushed posterosuperiorly and trachea was central. Thyroid scan revealed the thyroid tissue in the normal location. Patient's baseline laboratory investigations were within normal limits including serum amylase level. The surgical removal of the mass was planned. Keeping in view that it was fully occupying the oral cavity and lack of facility of fibroptic intubation, tracheostomy seemed to be the only option.

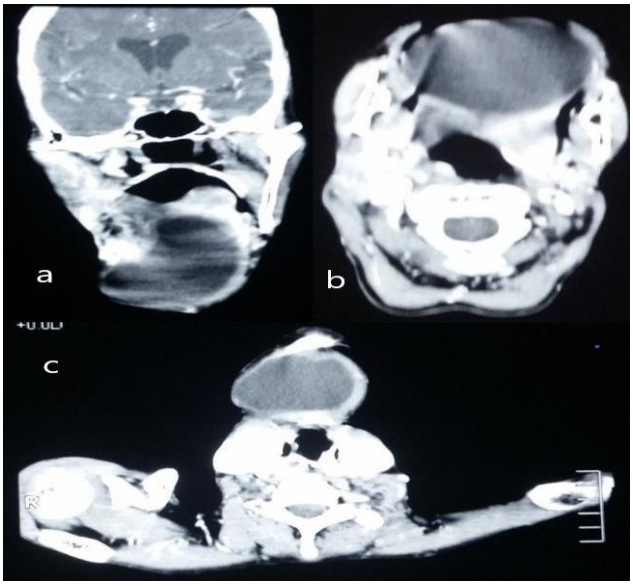

Figure 2: CT Scan with contrast showing a) Large minimally complex predominantly cystic lesion pushing tongue posterosuperiorly and measuring 69 mm transverse and 95 mm craniocaudal length and b) 74 mm anteroposterior dimension, filling the oral cavity and c) with significant extension into the neck. It has 6 mm thick wall.

After counseling about the nature of the lesion, surgical risks involved and the possibility of a surgical airway, pre-anesthetic evaluation was performed with special emphasis on the airway. The patients mouth opening was more than 3 finger, with normal flexion and extension of the neck, however with a large mass occupying the oral cavity and tongue pushed back into the pharynx (figure Ia), indirect laryngeal examination was not possible and the Mallampatti grade was IV which was a real challenge. CT scan did not show any laryngotracheal compression or deviation.

Keeping in view findings of preanesthetic checkup, it was mutually decided to attempt an endotracheal intubation using the skills of a senior anesthetist and with a surgical airway as last resort and the swelling was removed through trans cervical approach. In the operating room, with trolley prepared for difficult airway management including surgical airway, monitors were attached and the patient was preoxygenated with 100% oxygen. The patient was premedicated with 0.2 mg glycopyrrolate, 2 mg midazolam, and 4 mg Nalbuphine, followed by induction with Injection Propofol and ventilation. Inj.

Succinyl choline 50 mg was given and the patient was intubated on second attempts with No.6.5 cuffed endotracheal tube with the stylet and using Macintosh laryngoscope with a large blade. Excision was done by a Trans cervical approach (figure 4a) and a mass was successfully removed (figure 4b) without injury to neighboring structures. Post-operative recovery was uneventful. The diagnosis was supported by histopathological examination, which revealed a benign lesion, composed of large lymphatic channels lined by flattened endothelial cells, in loose connective tissue.

Stroma revealed foci of aggregates of lymphocytes, forming lymphoid follicles. No evidence of malignancy was noted. One-year follow-up (figure 5), did not reveal any complication or recurrence.

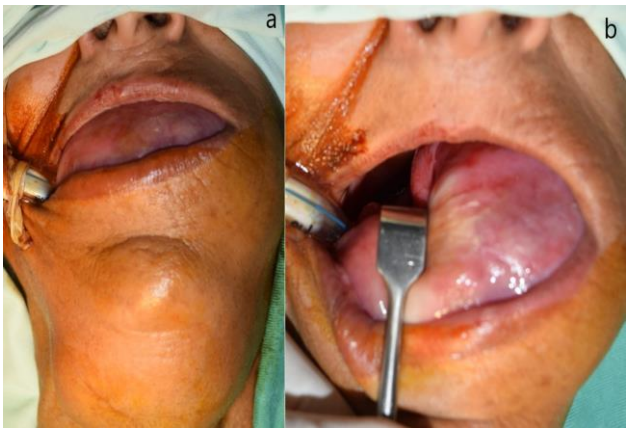

Figure 3: Preoperative Clinical photograph showing the available space in the oral cavity and Oro-tracheal tube in place.

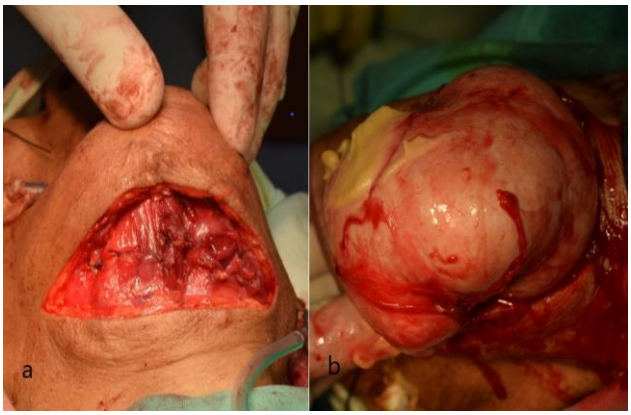

Figure 4: a) Trans cervical incision site at after removal of the mass and b) Large CH at the time of removal trans cervically.

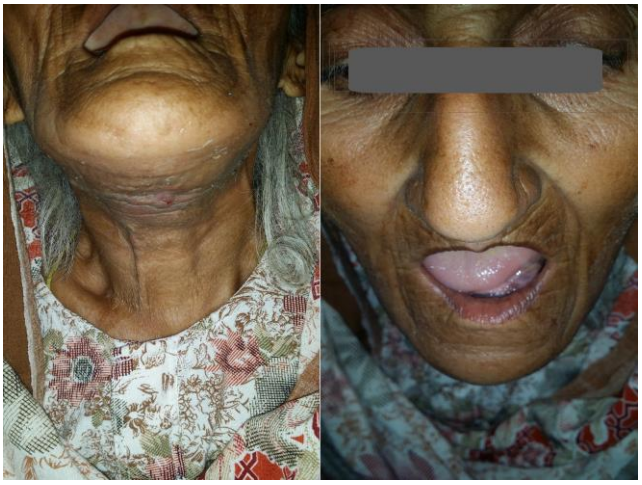

Figure 5: Photograph taken one-month post operatively showing scar of trans cervical incision and tongue back to the normal intraoral position with no residual swelling.

Discussion

Cystic hygroma is a benign congenital lymph angiomatous malformation with the incidence of 1.2 to 2.8 per 100,000 infants. [4,5] Of these, 50 to 60% are evident at birth and 80 to 90% present by 2 years of age.[4,5] Cystic hygroma are rare in adults with less than 150 cases reported in English language literature.[6,7,8] Etiology in adults is uncertain, although trauma, obstruction of lymphatic channels and infections have been implicated as possible triggers.[4] No gender predominance is noted.[9] Around 75% cases of CH occur in the Head and Neck region mostly affecting posterior triangle and 20% occur in the axilla followed by mediastinum and other sites.[1-3,10] Symptomatology depends on the anatomic location, size of the tumor and also the age of the patient. Obstructive symptoms are rare in adults, however, when occur they include dysphagia, dysphonia and dyspnea.[11] de Serres[12] proposed a staging system according to location and extent of lesions which labels cystic hygromas which are unilateral infrahyoid as stage 1, unilateral suprahyoid as stage 2, unilateral infrahyoid and suprahyoid as stage 3, bilateral infrahyoid as stage 4, and bilateral infrahyoid and suprahyoid as stage 5.[12] According to this classification, the present case with a large mass almost completely occupying the oral cavity as well as bilateral submandibular and submental extension stands at de Serres stage 5 (figure I).

Evaluation to visualize tumor extent is essential including extension into neck spaces and thoracic seen in 10%

cases.[13] Literature highlights role of preoperative imaging modalities like Ultrasonography, MRI, and CT [11] however, choice of modality usually depends on affordability, convenience, and resolution. MRI is the consensus study of choice since it provides remarkable soft tissue detail and relations to neighbouring structures.

In contrast CT scanning is rapid and readily available but lesion may not be easily delineated from surrounding soft tissues. The use of Fine needle aspiration cytology (FNAC) in cystic hygroma is controversial and it can result in infection, bleeding, and recurrence.

Morbidity associated with CH, depends on site, and impingement on critical structures like airway, nerves, vessels, and lymphatics. Mortality is usually due to airway compromise, accounting for 2-6% cases and usually encountered due to the extension of the CH in the oral cavity [10], this being the cause of difficult airway in our case. Other causes include intrathoracic extension, pressure on the larynx and trachea, co-existing congenital malformations, hemorrhage and post-operative respiratory obstruction. [14]

Airway management in cases of large CH's is a challenge for both the surgeon and the anesthetist. Therefore, assessment is essential in order to devise an appropriate airway plan for surgery and post-operative management. It necessitates making use of suitable imaging technique to assess the site, size and extensions of the tumor, pre-anesthetic check-up, airway assessment, preparation of trolley for difficult airway as well as intra operative and post-operative care[15], as done in our case. A well-equipped difficult airway trolley with all possible options for intubation and surgical airway and a standby surgeon to handle a can't intubate and /or can't ventilate situation. Blind nasal or oral intubation requires expertise but can result in injury and bleeding. This should only be attempted by a skilled senior anaesthetist as done in this case. Safer option is fiberoptic intubation, this too requires skills and experience, and however availability is an issue in most of our centers. Cricothyrotomy, tracheostomy may be difficult in lower neck swellings. Before surgery aspiration of the cyst can ease intubation, but can make surgery difficult.[14] CH's can be staged based on anatomical location to predict the outcome [12] and make a plan to establish and maintain airway for the successful surgical outcome. In our case of a large de Serres[12] stage 5 cystic hygroma occupying the whole of oral cavity with feeding and breathing difficulty, was intubated using a mackintosh laryngoscope by skilled senior anaesthetist with preparations for a surgical airway. 100% complication rate has been reported for de Serres stage 5 disease[12], our case had dysphagia and some breathing difficulty preoperatively, however, no post-operative complication occurred. Though usually, cystic hygroma with difficult intubation are reported in infants[6] only a few cases with difficult intubation have been reported in adulthood. Singh at al reported a rare case of huge cystic hygroma in an adult with difficult intubation.[16] Though Singh et al., faced problem due to huge neck mass, our case was different because of the mass occupying the oral cavity, which made airway difficult.

Among the cystic neck swellings in adults, CH is an uncommon differential diagnosis[17] and diagnosis presents a greater challenge with final diagnosis usually based on postoperative histology.[4] Likewise, in our case the FNAC was inconclusive and final diagnosis was based on postoperative histopathology.

Complete surgical removal is the preferred treatment option, others include laser surgery, cryotherapy, electrocautery, steroid administration, sclerotherapy, and embolization and radiation therapy.[3] Cystic hygroma varies from 1.0 to 30.0 cm in size; the mean size in Stromberg's series was 8.0 cm.[18]

The present case was unusual, with a large cervical cystic hygroma measuring 74 mm Anteroposterior, 69 mm transverse with 95 mm craniocaudal length on CT scan, presented de novo in a 61-year-old lady with no history of trauma or upper respiratory tract infection, with difficult intubation. Total surgical excision was possible after a difficult intubation by the skilled experienced anaesthetist.

Conclusion

Cystic hygroma may rarely be encountered in adulthood, when dealing with cystic cervicofacial masses and should be considered in the differential diagnosis. In such cases a difficult airway may present a challenge. Morbidity and mortality can be avoided by proper airway management and complete surgical excision in experienced hand. An experienced skilled anesthetist and all preparations to handle a difficult airway are essential.

Acknowledgement

The work of Dr. Syed Arshad Ali, Anesthetist is acknowledged, for in helping us out with such difficult cases.

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