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CYSTIC HYGROMA CO-EXISTING WITH CONGENITAL SUBGLOTTIC STENOSIS

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Case report
ABSTRACT
Respiratory distress is one of the commonest cause of admission into a Neonatal Intensive Care Unit, be it surgical or medical in nature. Adequate and prompt resuscitation as well as intubation can be life saving. Emergency or early tracheostomy may be necessary if airway intervention is needed. The authors present the case of a term neonate who was born with a large cervical cystic hygroma causing compression of the airway, together with a concurrent grade III subglottic stenosis, in respiratory distress within few minutes of life. Nine months post tracheostomy and sclerotherapy treatment twice, the child shows marked improvement. The succesful management of this unusual case of severe extrinsic compression with concurrent internal airway obstruction is presented.
Key words: Sublottic stenosis; Cystic hygroma; Respiratory distress.

1. INTRODUCTION

Cystic hygroma is a form of lymphangioma. Large enough, it has the capability of causing airway compromise due to external mass effect. Subglottic stenosis on the other hand, is the third most common congenital laryngeal abnormality making up 15% of all congenital laryngeal abnormality cases (1). However, only 5% of cases are of the congenital type, as opposed to its rather common acquired counterpart. These two separate entities presenting simultaneously in the same patient is extremely rare, and can result in undesirable complications if treated inadequately. We report such a case in a neonate presenting with respiratory distress.

2. CASE REPORT

A term baby weighing 3.0 kg was born via emergency lower segment caesarean section for foetal anomaly in labour. A large cervical cystic hygroma was detected prehand during antenatal ultrasonography. Within 10 minutes of life, the child could not sustain her saturation. She developed stridor and went into respiratory distress. Prompt resuscitation was commenced, however multiple attempts at intubation with endotracheal tubes size 3.5 and 3.0 failed. Finally she was intubated successfully with a 2.5 sized tube and ventilated in the neonatal intensive care unit.

Clinically she was noted to have a soft, cystic, transilluminating, multiloculated swelling measuring about 80 x 60 mm. The cyst extended from the lateral aspect of the right side of the neck, i.e from the right tragus, up to the anterior aspect of the neck, crossing the midline, causing the head to be rotated to the left side (Figure 1). No syndromic features were noted as such. Computed Tomographic scan showed a multilobulated, heterogenous, non-enhancing multiseptated cystic mass measuring 86 x 43 x 58 mm (Figure 2). The airway was narrowed and displaced to the left. The trachea however was noted to be patent at the level of the thoracic inlet.

Intravenous Dexamethasone was started at 0.25mg/kg/hour and we proceeded with direct laryngoscopy and tracheostomy under general anaesthesia on Day 6 of life. On direct laryngoscopy, bulging of the external mass was noted internally. The epiglottis, glottis and arytenoids were oedematous. The subglottis was found to be stenosed anteriorly, with about 10% airway patency posteriorly (Cotton Grade III)

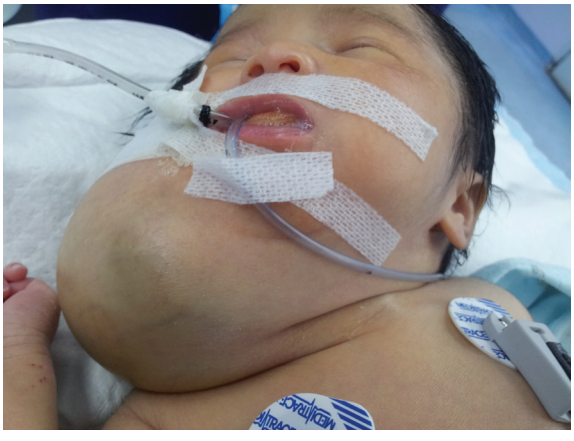

Figure 1. Large cystic sweling on the right side of the neck.

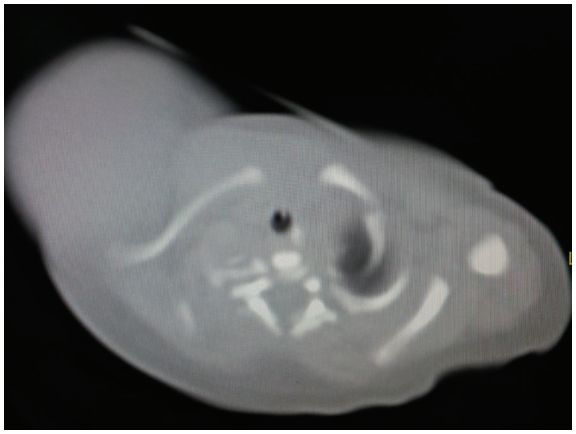

Figure 2. CT scan demonstrating a huge non-enhancing lesion.

(Figure 3). The trachea was deviated to the left, hence it was approached medial to the cyst for the tracheostomy. A non-cuffed tracheostomy tube size 3.5 was inserted uneventfully. The child currently 9 months old, still on a tracheostomy tube, is active and feeding well. The mass, post scleotherapy twice, has significantly reduced in size.

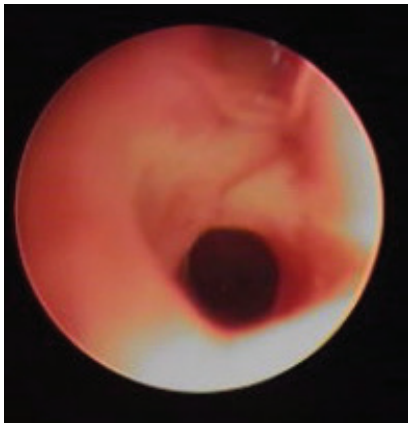

Figure 3. Grade III subglottic stenosis as evident on direct laryngoscopy

3. DISCUSSION

Cystic hygromas are the macrocystic type oflymphangioma containing large cysts, and occuring due to lymphatic malformation. They may be congenital or acquired. They occur anywhere in the human body, with a prediliction for the head and neck region (75% of cases) (2). Management of these cysts may be observational or in larger symptomatic cases, sclerotherapy with agents such as bleomycin and ethanol may be advocated. In view of the fact that intrauterine cystic hygromas have been associated with Turner's Syndrome (3), Down and Noonan Syndromein previous literature, cytogenetic studies in such cases are warranted.

Subglottic stenosis occurs due incomplete recanalization of the laryngotracheal tube. They can be classified into the cartilaginous or the membranous type. The smaller stenosis may not manifest until a child develops stridor due to a respiratory tract infection. On the other end of the spectrum, the more severe ones may be evident with early respiratory distress, as in this case. Direct laryngoscopy and tracheobonchoscopy is the gold standard for diagnosis.

The management of a subglottic stenosis may be conservative or surgical. Most stenoses tend to resolve as the child grows older and most often, children on a tracheostomy may be decannulated by the age of 3. However in the more severe cases, surgical approach such as an anterior cricoid split, laryngotracheal expansion surgery or cricotracheal resection may be advocated. The main aim is to establish a sufficient airway without the need for a tracheostomy, a good cough reflex and a fair voice quality. These surgeries however, are not without their own complications.

The child in this particular case had a double jeopardy causing severe airway obstruction. The presence of a large neck mass compressing on the trachea and shifting it to the contralateral side made intubation a challenge to begin with. Furthermore, securing the airway was not made any easier with an internal narrowing due to the significant subglottic stenosis. Hence, the judicious decision for an early tracheostomy was made after much deliberation in view of the fact that this was a case of an extremely difficult intubation, and should the endotracheal tube dislodge, it would be an uphill task to reinsert it back. The technical difficulty and challenge posed by the distorted anatomy, and the possible risk of a tracheo-arterial fistula formation was born in mind pre-operatively.

The occurence of a dual pathology, i.e congenital cystic hygroma together with a subglottic stenosis simultaneously in the same patient is rare. It may be attributed to co-incidence or there could be some form of genetic association. Such associations of these two pathologies have not been described in literature thus far. As for the further management of the stenosis, it is yet to be decided till the child grows older. Only then can she be reassessed and managed accordingly; be it conservatively or surgically.

4. CONCLUSION

Although congenital cystic hygromas and congenital subglottic stenosis may be self-limiting in their respective manners, the simultaneous occurence of these pathologies in its severe form can be lethal if inadequately identified and treated. It might be worthwhile to bear in mind the presence of a congenital subglottic stenosis in a child with congenital cystic hygroma in the future.

Conflict of interest: none declared

REFERENCES

1. Manoukian JJ, Tan AK. Embryology of the larynx. In: Tewfik TL, Der Kaloustian VM, eds. Congenital Anomalies of the Ear, Nose, and Throat. New York, NY: Oxford University Press; 1997: 377-382.
2. Zainine R, El Aoud C, Sellami M, Beltaief N et al. Cystic hygroma: report of 25 cases. Tunis Med. 2012 Jan; 90(1): 19-24.
3. Kassanos D, Botsis D, Pyrgiotis E, Salamalekis E. et al. Cystic hygromas associated with Turner's syndrome. Report of three cases. Clin Exp Obstet Gynecol. 1992; 19(4): 222-225.
4. Altman KW, Wetmore RF, Marsh RR. Congenital airway abnormalities in patients requiring hospitalization. Arch Otolaryngol Head Neck Surg. May 1999; 125(5): 525-528.