Congenital vallecular cyst with polydactyly- A rare cause of stridor and failure to thrive

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SUMMARY

Congenital vallecular cyst is an uncommon, benign but potentially dangerous condition causing respiratory distress and stridor. It is associated with sudden upper airway obstruction resulting in death due to its anatomical location in neonates and infants. We reported a rare case of 2 months old male infant presented with respiratory obstruction with failure to thrive with polydactyly (rarest finding) and managed timely with appropriate surgical marsupialization.

KEY WORDS:
Congenital vallecular cyst, Failure to thrive, Polydactyly

INTRODUCTION

Congenital vallecular cyst (VC) is an uncommon but potentially life threatening cause of respiratory distress, stridor and even death[1]. VC is also named as epiglottic cyst, mucous retention cyst, ductal cyst and tongue base cyst[2]. Feeding difficulties, coughing, voice changes, dyspnea, stridor and failure to thrive are the most common symptoms[3]. VC has prone to excite the retching reflex which induces gagging and vomiting. These feeding difficulties may leads to failure to thrive[1]. Early detection and timely definitive therapy of VC reduces the morbidity and mortality since the initial presentation of it often mimics common benign causes of neonatal airway obstruction which is easily overlooked. Developing country like India with poor health setup at rural level, a significant portion of VCs continues to be identified only at autopsy. Our primary aim is to improve clinical awareness of congenital VC through an illustration of our departmental recent experience with this condition. Furthermore, we explored the current management of neonatal stridor caused by this devastating congenital condition.

CASE REPORT

8 months and 5 days gestational age male baby (weight- 2.5 Kg) has been born by normal vaginal delivery without any perinatal problems. On day of life 7, baby developed difficulty in respiration, noisy breathing and recurrent vomiting. He was brought to local doctor and treated for lower respiratory tract infection (LRTI) with injectable antibiotics and nebulization. Symptoms had persisted at 2 months and 5 days of age. He was referred and admitted to our department for management of progressively developing stridor.

At 2 months and 12 days old, baby weight (2.75 Kg), height 48 cms (both < 3rd percentile) and head circumference was 36cms (anthropometry suggestive of failure to thrive). Physical examination revealed respiratory rate were 54/minutes, heart rate 146/minutes. Muscle bulk was hypovolemic but strength and power were normal. On general examination, no rash or petechiae on skin but incidentally, he has an extra 6th finger on his right thumb (see Figure 1). Oral examination showed "sun rising" cystic lesion at the base tongue while ear and nose examination were normal.

Normal Chest X- ray, USG abdomen normal (to rule out any anomalies) and 2DECHO were also normal. CT scan showed well defined clear enhancing mid-line thin wall
cystic lesion of 16x15mm at the base tongue indenting oropharynx (See CT photograph). Thyroid gland, trachea, larynx and lower airway appeared normal.

Under guidance of video-laryngoscope, cyst were aspirated partially and again filled with methylene blue (to define cyst wall). The marsupialization of cyst was performed under general anesthesia.

Post operatively, naso-gastric feeding were given for 3 days and graded to oral feed. Patient's symptoms immediately resolved. Rechecked video-laryngoscopy on day seven showed no evidence of recurrence with small raw area at the base tongue. Baby was discharged healthy with no airway and feeding problem and gained weight steadily.

DISCUSSION

Benign laryngeal cysts, a rare inhomogeneous group of airway abnormalities. The annual incidence is 1 per $3x10^5$ and 1.82 per 10^5 births in selected population. The most common laryngeal cyst in the newborn is aryepiglottic cyst followed by VC, ventricular cyst and subglottic cyst. In literature, VC comprises 10.5% of all congenital laryngeal cysts. Still the precise incidence of VC is not known, it is regarded as extremely rare, especially in its congenital form[4].

VC is unilocular cystic mass of variable size containing clear and non-infected fluid. Hypothetically, VC is a consequence

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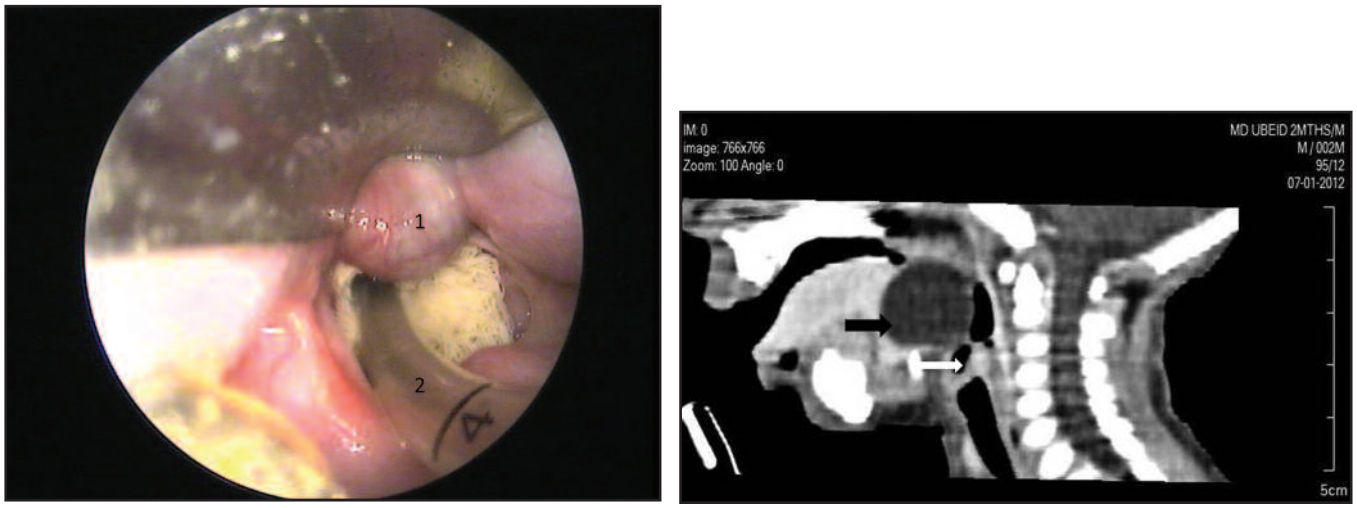

Fig. 1 : Intraoperative photograph of 2 month old male baby with vallecular cyst (Label- 1). Label -2- endotracheal intubation. CT PHOTOGRAPH: CT (Base skull to thoracic inlet) showed a well-defined clear enhancing mid-line thin wall cystic lesion of 16x15mm at the base tongue (Black Arrow) (White Arrow- Epiglottis).

of either ductal obstruction of mucous gland or an embryological malformation. Histologically, VC lined by respiratory epithelium with mucous gland internally while an external lining of squamous epithelium[1].

Abercrombie described the first laryngeal cyst in 1881[5]. Wood R. E. (1984) studied 225 children with stridor and noted that 4 children had supraglottic cysts and 2 had subglottic cysts but there were no vallecular cyst in this study[6]. A largest series conducted by Mitchell *et al* in 1987, over period of 15 years, not a single case of VC were noted[5]. Published study by Hsieh and Liu were diagnosed 11 and 14 cases of congenital VC respectively[7].

VC is a potentially devastating cause of upper airway obstruction with high morbidity and mortality in newborn and infants owing to its anatomical location in combination with relatively small size paediatric airway. Such airway obstruction caused by mass effect in hypopharynx and significantly postero-inferior displacement of epiglottis which leads to sudden supraglottic obstruction[4].

Usually the Infant with VC have symptoms during first week of life. Infants with VC may present a secondary form of laryngomalacia because due to altered airway dynamics caused by progressively enlarging VC may elevate inspiratory negative pressures contributing to supraglottic prolapsed and a secondary from of laryngomalacia developed. Failure to thrive reported in approximately 66% cases which are caused by combined effect of laryngomalacia, gastro-oesophageal reflux in infant with VC[1]. Our case presented with above mentioned combined effect with right polydactyly.

In the differential diagnosis of VC, thyroglossal duct cyst, dermoid cyst, hamartoma, teratoma, cystic hygroma and hemangioma should be considered. Surgical treatment for VC like complete surgical excision, endoscopic marsupialization (by using CO_2 LASER) and Deroofing of cyst. We performed endoscopic marsupialization by cold dissection method under guidance of methylene blue staining of cystic wall which is safe and definitive procedure. Simple cystic aspiration is not recommended because of its high recurrence rate[1]. The major complication of surgery includes accidental rupture of cyst and aspiration, incomplete marsupialization and endotracheal intubation induced hemorrhage into the cyst[4].

CONCLUSION

Vallecular cyst in newborn and infant is a serious and life threatening situation that require special attention of parents and health care providers at rural level. History of progressive respiratory distress and failure to thrive makes its necessary that the treating paediatrician must always keep the congenital cause of upper airway obstruction in mind when dealing with a respiratory case. The combination of detail history, clinical signs and radiological signs are more specific than each one separately. Timely management of such devastating condition gives some assurance of survival in the future.

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