CASE REPORT

Congenital Vallecular Cyst: A Rare and Potentially Lethal Condition

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ABSTRACT

Congenital vallecular cyst is a rare laryngeal lesion, accounting for 10-20% of all laryngeal cysts, with a potential to cause severe upper airway obstruction, which can be fatal. It can cause stridor, apnea, cyanosis, respiratory distress, and feeding difficulties. Diagnosis requires a high level of clinical suspicion and helps in timely intervention. Direct laryngoscopy is gold standard for definitive diagnosis. Treatment options include aspiration, marsupialization, and surgical excision. Here, we report a case of 7-week-old infant with complain of recurrent episodes of cyanosis when agitated, since the age of 3 weeks, admitted with impression of apparent life-threatening events (ALTEs). Extensive investigations were non-conclusive, which were done in local hospital. Direct laryngoscopy was performed in our hospital and showed presence of a vallecular cyst. Thus complete excision of cyst was done in the same setting with dramatic relief of symptoms.

Key Words: Congenital vallelar cyst. Cyanosis. Respiratory distress.

INTRODUCTION

Vallecular cyst is a rare, but potentially lethal cause of upper airway obstruction in infants and young children. It usually presents with stridor, respiratory distress, and feeding difficulties; but can lead to death, caused by supraglottic obstruction due to mass effect.1-2 The precise incidence is unknown.

Vallecular cyst arises from the lingual surface of the epiglottis. It occurs as a result of either ductal obstruction of mucous glands or an embryologic malformation. Direct laryngoscopy remains the best diagnostic tool.3,4 Definitive treatment options include aspiration, marsupialization or complete excision of the cyst.5

We describe a case of vallecular cyst in a 7-week-old infant admitted in our hospital, initially seen by general pediatrician and then referred to ENT surgeon for further management.

CASE REPORT

A 7-week-old infant admitted in Child Health Department (CHD) of Sultan Qaboos University Hospital (SQUH), through Accident and Emergency Department (AED) with impression of apparent life-threatening events (ALTEs). She was born full-term by spontaneous vaginal delivery with no perinatal and postnatal complication. She was doing well till the age of 2 weeks. Then, she presented in a local hospital with history of recurrent episodes of apnea and cyanosis provoked by excessive crying; associated with respiratory distress, cyanosis, and desaturation upto 70 - 80% lasting for few minutes and resolved spontaneously. On examination, she was vitally stable. There was no dysmorphism and systemic examination was unremarkable. Extensive work-up was done including CT brain, found to be normal except for hypochromic microcytic anemia with hemoglobin of 8 gm/dl and she was transfused with packed red blood cells (PRBCs) and investigated for the cause and discharged home. Again at the age of 7 weeks, she presented with the similar episode of cyanosis, respiratory distress and desaturation with excessive crying, but this episode was more prolonged and lasted for about 1 hour and she was transferred from the local hospital to SQUH for further evaluation.

During hospitalization, while drawing the blood sample, she developed cyanosis, respiratory distress manifested by sternal, suprasternal, subcostal and intercostal recessions with obvious inspiratory stridor and desaturation up to 70% in room air, improving to 100% with oxygen support.

In between the episodes, the child was vitally stable on apnea monitor, maintaining saturations in room air and comfortable in mother's lap with no cyanosis, stridor, and respiratory distress. Extensive work-up was inconclusive, ECHO was normal. Respiratory viral screen was positive for influenza B and rhinovirus. ENT review was taken as this apnea was not explained by the viral infection only. They tried to assess the patient at bedside by flexible laryngoscopy, but she had an episode of desaturation. Direct laryngobronchoscopy was performed under general anaesthesia, which showed a cystic mass at the base of tongue and vallecula pushing the posterior pharyngeal wall and obstructing the
Congenital vallecular cyst is a rare benign lesion which commonly arises from the lingual surface of the epiglottic region, and is classified as ductal cyst that results from obstruction and retention of mucus in collecting ducts of submucosal glands containing clear and non-infected fluid.5 Neonates usually present with signs and symptoms of upper airway obstruction, which warrants further evaluation and investigation. Common presenting features are inspiratory stridor, respiratory distress, apnea, cyanosis, and hoarse cry. These patients may present with acute, life-threatening, airway compromise, requiring emergency airway management. Fatal or near fatal cases have also been reported. Majority of affected infants presents in the first few weeks of life with median age of 3 and 40 days at diagnosis.1 This cyst has also been reported in older children and adults, in whom it is usually detected during the intubation for the procedures. Older children can present with feeding difficulties and failure to thrive.

The clinical presentation may further complicate with coexistent laryngomalacia, which influences management plan. In this case, there was no evidence of laryngomalacia and the symptom was completely resolved after surgical excision.

Imaging studies, like ultrasound, computed tomography and magnetic resonance imaging (MRI), help in the diagnosis of vallecular cysts. MRI is a valuable imaging method in determining the extent and contents of the lesion. Prenatal detection with ultrasound or MRI is helpful for immediate action and excision of the cyst postnatally.5 However, in cases with high clinical suspicions of upper airway obstruction, flexible laryngoscopy is recommended for the initial screening and definitive diagnosis. Management of vallecular cyst includes aspiration of cyst, marsupialization or complete excision. Surgical excision is the treatment of choice.4 Long-term prognosis is generally good with minimal recurrence risk.6

Signs and symptoms of severe respiratory distress in a neonate due to upper airway obstruction warrant early diagnosis and immediate action. Differential diagnosis includes laryngomalacia, cystic hygroma, lymphangioma, hemangioma, dermoid cyst, thyroid remnant cyst, and thyroglossal cyst. Doctors have to act immediately, using the clinical suspicion. Further evaluation by history and clinical examination will narrow the list of differential diagnosis and will help in minimizing the workup.

As in our case, imaging studies were not done and clinical suspicion was enough to direct for focused investigations and appropriate management.

In the literature, no local data is available for this condition.

REFERENCES