BRIEF COMMUNICATION

Congenital Saccular Cyst of the Larynx: A Rare Cause of Stridor in Neonates And Infants

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Abstract Stridor is a noise caused by the passage of turbulent air through a diminished airway caliber. Laryngomalacia is the most common congenital anomaly of the larynx and the principal cause of stridor in children under 6 months.

A less common aetiology of stridor and respiratory distress in newborns and infants is congenital laryngeal saccular cyst. This entity should be considered in the differential diagnosis of stridor. Early recognition and proper treatment are essential because it can cause life-threatening airway obstruction.

We describe our experience with 4 patients with this disease, 3 of them were successfully treated with microsurgical resection of the cyst, and we emphasise the importance of considering the endoscopic evaluation of the airway in every child exhibiting stridor with an unexpected evolution to determine the causal lesion.

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PALABRAS CLAVE

Estridor; Quiste laríngeo congénito; Niños; Microcirugía laríngea

Quiste saccular congénito de laringe: una causa rara de estridor en neonatos y lactantes

Resumen El estridor es un ruido ocasional por el pasaje de aire en forma turbulenta mediante una vía aérea con calibre disminuido. La laringomalacia es la anomalía congénita más común de la laringe y la principal causa de estridor en los menores de 6 meses.

Una etiología menos frecuente de estridor y dificultad respiratoria en el recién nacido y lactante es el quiste saccular laríngeo congénito, y debe ser considerado en el diagnóstico diferencial del estridor. El reconocimiento precoz y el tratamiento adecuado son esenciales ya que puede causar obstrucción de la vía aérea con amenaza de vida.


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Describimos nuestra experiencia con 4 pacientes que presentaban esta afección, 3 de ellos tratados exitosamente con resección microquirúrgica del quiste, y destacamos la importancia de considerar la evaluación endoscópica de la vía aérea en todo niño con estridor cuya evolución no es la esperada para determinar la lesión causal. © 2012 Elsevier España, S.L. Todos los derechos reservados.

Introduction

Stridor is a noise produced by an alteration in the airflow passing through a partially obstructed airway. The main cause in newborns and infants is laryngomalacia.1,2

Saccular cysts (SC) are unusual congenital lesions which clinically appear in the neonatal period or early infancy.3 They develop at the supraglottic level by a blockage of the opening of the saccule of the laryngeal ventricle. Their incidence is 1.82 cases per 100,000 live births.2 The most common symptom is inspiratory stridor and the initial presentation may mimic laryngomalacia. As the cyst grows, it is associated with progressive respiratory distress, episodes of cyanosis and apnoea, hoarse or low crying and difficulty in feeding.

The diagnosis is confirmed by endoscopic visualisation of the lesion and cervical computed tomography (CT) scan, which will define its exact location and extension.4 Treatment is surgical, with various possible endoscopic techniques and external approaches. They may recur and may occasionally require a temporary tracheotomy.5

Case Reports

We report 4 patients with congenital SC who attended consultation at the Respiratory Endoscopy Service (referral centre for paediatric airway involvement) due to stridor and/or progressive respiratory distress from birth, between February 2010 and March 2012 (Table 1).

<table>
<thead>
<tr>
<th>Patient</th>
<th>Gender</th>
<th>Symptoms</th>
<th>Endoscopy</th>
<th>Age at diagnosis</th>
<th>SAPS</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>ML</td>
<td>F</td>
<td>Inspiratory stridor, hoarse crying, difficulty for breathing and feeding</td>
<td>Right lateral saccular cyst</td>
<td>6 months</td>
<td>–</td>
<td>Transoral puncture of the cyst, and LMS with CO₂ laser after 15 days</td>
</tr>
<tr>
<td>MM</td>
<td>F</td>
<td>Inspiratory stridor, difficulty for breathing and feeding and cyanosis</td>
<td>Left lateral saccular cyst</td>
<td>15 days</td>
<td>Otracheal intubation</td>
<td>Transoral puncture of the cyst, and LMS with CO₂ laser after 8 days</td>
</tr>
<tr>
<td>LJ</td>
<td>F</td>
<td>Inspiratory stridor, hoarse crying and progressive difficulty for breathing</td>
<td>Right lateral saccular cyst</td>
<td>11 months</td>
<td>–</td>
<td>LMS with CO₂ laser</td>
</tr>
<tr>
<td>BA</td>
<td>M</td>
<td>Inspiratory stridor</td>
<td>Left lateral saccular cyst</td>
<td>15 days</td>
<td>–</td>
<td>Waiting attitude at 3 moths follow-up</td>
</tr>
</tbody>
</table>

F: female; LMS: laryngeal microsurgery; M: male; SAPS: stabilisation of airway prior to surgery.
Laryngeal microsurgery was performed after 1 month (Fig. 2). Patient 4, diagnosed at 15 days of life through fibro-laryngoscopy and CT, has not yet undergone microsurgery since he does not suffer difficulty for breathing (Fig. 3).

Operated patients were extubated in the operating room, fed orally within 24 h and received postoperative treatment with intravenous antibiotics and steroids. They were discharged between 3 and 5 days after surgery.
The anatomopathological study reported that each cyst wall was lined with respiratory type epithelium and did not present cellular atypia, thus supporting the diagnosis of SC. No complications were observed. All 3 patients presented excellent immediate evolution, with spontaneous ventilation, no audible stridor or dysphonia and no difficulties for swallowing. They currently remain asymptomatic after 3, 16 and 28 months monitoring, respectively.

Discussion

Congenital laryngeal cysts may be saccular or ductal. Ductal cysts represent 75% of all congenital laryngeal cysts. They originate from an obstruction of the excretory duct of laryngeal epithelial mucous glands and are mainly located in the vallecula. They usually appear later in life than SC, causing difficulty in swallowing. Congenital SC are a dilatation with mucous content of the saccule of the laryngeal vestibule, resulting from a developmental failure to maintain patency of the opening orifice. The saccule extends vertically between the false vocal cord, base of the epiglottis and inner surface of the thyroid cartilage. It contains mucous glands whose secretions lubricate the vocal cords.

There are 2 types of SC: anterior, which project into the anterior region of the ventricle and protrude into the laryngeal lumen between the true and false vocal cord, and lateral, which are larger and more frequent and which appear as a bulge of the ventricular band and/or arytenoepiglottic fold.

SC represent 1.5% of congenital laryngeal anomalies. Although rare, they should be included in the differential diagnosis of stridor and progressive obstruction of the airway in neonates and infants. Since the diagnosis of laryngomalacia is by far the most common, paediatricians tend to think of this entity in most cases of stridor, without conducting a nasofibrolaryngoscopic assessment of the patient, thus leading to errors when the real aetiology is another.

Stridor at birth is rare and is usually due to a congenital narrowing of the subglottis (stenosis or membrane). Dynamic conditions, such as laryngomalacia or vocal cord paralysis, usually become evident during the first week of life. SC typically appear with inspiratory stridor and may be mistaken with laryngomalacia. When caused by SC, stridor may be present from birth. As the size of the cyst increases, stridor may also increase and be associated with voice changes (perceived as hoarse or low crying), episodes of cyanosis, retraction, and difficulty for feeding, even reaching a complete obstruction of the airway. On the other hand, laryngomalacia never causes changes in the quality of voice. Another difference is that, in laryngomalacia, stridor decreases in the prone position, whereas in lateral SC, symptoms decrease when the patient is lying on the affected side and increase in the contralateral position.

The diagnosis is suspected through the symptoms and clinical course. Imaging tests and fibrolaryngoscopy may also point in that direction initially. Although lateral neck radiographs reveal most cysts, CT and MRI scans are preferable because they offer a better definition of the lesion in terms of size, location, extent and anatomical relationships. Direct laryngoscopy under general anaesthesia with intubation secures the airway and confirms the diagnosis. Complete endoscopic evaluation of the airway (from nostril to bronchi) identifies various concomitant abnormalities, with the association of 2 or more lesions being identified in 10%-45% of patients with congenital stridor.

The differential diagnosis of SC should include ductal cysts, laryngocele, haemangioma, hamartoma and teratoma, among others. Due to their rarity, it is difficult to determine the optimal surgical strategy. Treatment includes securing the airway, usually with an endotracheal tube or even with tracheostomy in some cases, and cyst removal. The endoscopic approach is the most common choice.

Needle aspiration of the cyst may confirm the diagnosis and occasionally obviate the need for tracheostomy and/or a more definitive treatment. Endoscopic excision is recommended when the cyst recurs after puncture. Partial removal of the cyst wall or marsupialisation is rarely sufficient, with complete removal of the cyst wall being required to prevent recurrences. The external approach is reserved for recurrences after endoscopic treatment or in cases of extensive injury. The application of CO2 laser has been introduced for the excision or vaporisation of the cyst wall. Microsurgical resection of the cyst with CO2 laser proved to be a safe and effective procedure in our patients, with excellent results and without the need for tracheostomy or external incisions.

Obtaining an accurate diagnosis when the SC is the cause of stridor may be complicated, due to the inherent difficulty in paediatric examinations of the larynx. Moreover, being a rare entity, it is not generally considered during the evaluation of children with congenital stridor. Early diagnosis requires a high level of suspicion and is of great importance in order to reduce the morbidity and mortality associated with airway obstruction.

References