Indian J Otolaryngol Head Neck Surg
(Jan–Mar 2016) 68(1):118–122; DOI 10.1007/s12070-015-0960-2

CLINICAL REPORT

Congenital Bilateral Saccular Cysts and Bifid Epiglottis: Presentation and Management

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Received: 25 October 2015 / Accepted: 11 December 2015 / Published online: 19 December 2015

Abstract Saccular disorders are rare representing only 1.5 % of all laryngeal anomalies. Bifid epiglottis is also an extremely rare congenital anomaly that usually occurs in a syndromic picture in association with other anomalies such as polydactyly, cleft palate and micrognathia, which are seen in Pallister–Hall Syndrome and rarely with other syndromes. We report a case of bilateral saccular cysts and bifid epiglottis in a full term neonate presenting with stridor. The patient's other congenital anomalies included microretrognathia, short neck, polydactyly of four extremities and hypospadias. The patient underwent staged endoscopic microsurgical marsupialization of both cysts and endoscopic repair of the bifid epiglottis.

Keywords Stridor · Saccular cyst · Laryngocele · Bifid epiglottis

Introduction

Neonatal stridor can be caused by a variety of congenital and acquired causes. The most common congenital anomalies of the larynx with such a presentation include laryngomalacia (60 %), vocal cord paralysis (VCP) (10 %), and subglottic stenosis. Another rare cause is that of saccular disorders which almost always present with stridor [1–3].

Saccular disorders are secondary to abnormal dilatation of the saccule and include both laryngoceles and saccular cysts. They are classified according to the content and location. Depending on whether it contains air, mucus or purulent material, the lesion is named either a laryngocele with a patent saccular orifice, a saccular cyst with a blocked orifice or a laryngopyocele respectively. Saccular cysts are extremely rare representing only 1.5 % of all laryngeal anomalies [3, 4].

Bifid epiglottis is also an extremely rare congenital anomaly that occurs in a syndromic picture. Associated anomalies include polydactyly, cleft-palate, retro/micrognathia and asymptomatic laryngeal malformations seen in Pallister–Hall Syndrome (PHS) and rarely in other syndromes [5–7].

In this article we report a case of bilateral saccular cysts and bifid epiglottis in a full term neonate presenting with stridor and subsequent management. A brief review on the classification of saccular disorders, anatomy, clinical picture and the management is presented.

Case Report

We report a case of a full term male neonate who was the product of an uncomplicated pregnancy requiring a cesarean section due to premature rupture of the membranes. The Apagar score was 7 and 9 after 1 and 5 min respectively. The patient then started to have stridor and increased work of breathing with desaturation. He required intubation to secure his airway and was admitted to the neonatal intensive care unit. The patient had a prolonged

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stay and required tracheotomy. No bronchoscopy was done at that time. He was discharged with an appointment with the pediatric Otolaryngology clinic. During the time of his outpatient visit the patient was found to be vitally stable with no signs of respiratory distress or cyanosis. No neck masses were detected on physical examination. He did however have dysmorphic features including microretrognathia, short neck, hypospadias, post-axial right hand polysyndactyly, postaxial left hand polydactyly with extra tag and polydactyly of both feet. His thumbs, however, were normal (Fig. 1) Fiberoptic laryngoscopy revealed swelling of both aryepiglottic folds and a bifid epiglottis.

The patient's past medical history was unremarkable except for three previous episodes of seizures. The patient was sent for Ophthalmological and Genetic assessment. Laboratory investigations were normal. Blood gases showed persistent metabolic acidosis with normal anion gap. Computed tomography of the neck soft tissue confirmed the presence of two homogenous smooth walled cystic structures within the larynx. They fill bilateral pyriform sinuses with medial displacement of bilateral aryepiglottic folds. A normal appearing epiglottis could not be visualized (Fig. 2). Magnetic Resonance Imaging of the brain did not reveal any intracranial abnormalities. Specifically there was no evidence of a hypothalamic hamartoma. Barium swallow showed contrast penetration just superior to the vocal cords. Ophthalmologic assessment was normal. On genetic assessment the chromosomal analysis was found to be normal but the patient did have a positive history of consanguinity (parents are double first cousins). No similar conditions were in the family.

Fig. 1 Polysyndactyly of right hand

The patient was taken to the operating room and under general anesthesia ventilation was carried out through tracheostomy tube. Brnonchoscopy was performed and demonstrated complete laryngeal inlet obstruction by the presence of bilateral saccular cysts; a left large cyst and a smaller right. The epiglottis was found to be bifid. The subglottic area, trachea and bronchi were normal. Subsequently, suspension microlaryngoscopy was performed and staged removal of the cysts was planned. First to marsupialize the larger left cyst and to leave the other cyst for 4–6 weeks. CO_2 Laser was used to incise the cyst and mucoid thick fluid was suctioned from it. The wall of the cyst in the aryepiglottic fold causing obstruction of the piriform fossa was marsupilized and removed completely. The remaining edge was cauterized with laser. Hemostasis was secured. One Month later the right cyst was marsupilized in a similar fashion (Fig. 3).

Histologic examination of both cysts revealed fibrocollagenous tissue lined by respiratory-epithelium (pseudostratified epithelium) with no evidence of malignancy. Four weeks later, barium swallow was performed and showed aspiration of liquids at the level above the vocal cords. Decision was made to repair the epiglottis as bifidity was contributing to the aspiration. Suspension microlaryngoscopy was performed to endoscopically repair the

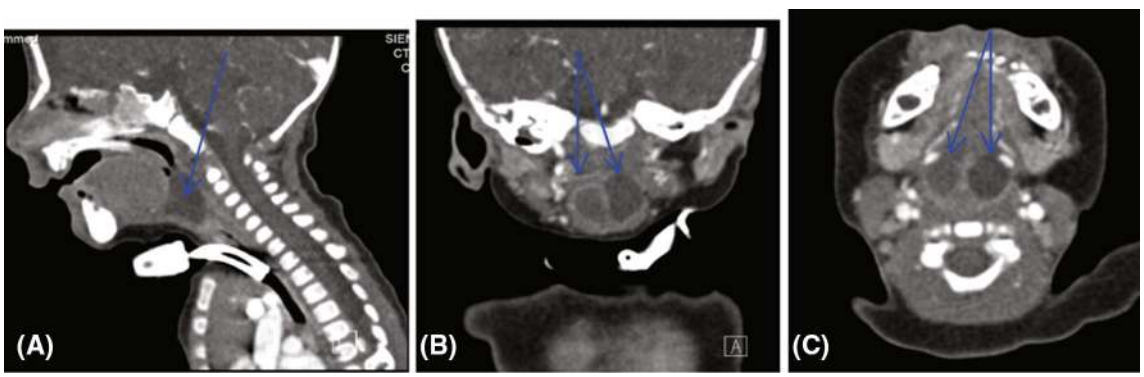

Fig. 2 CT scan—**a** Sagittal, **b** Coronal and **c** Axial images of the neck demonstrate the presence of two smooth walled cystic structures (*blue arrows*) within the larynx. The intervening soft tissue represents the medialized aryepiglottic folds

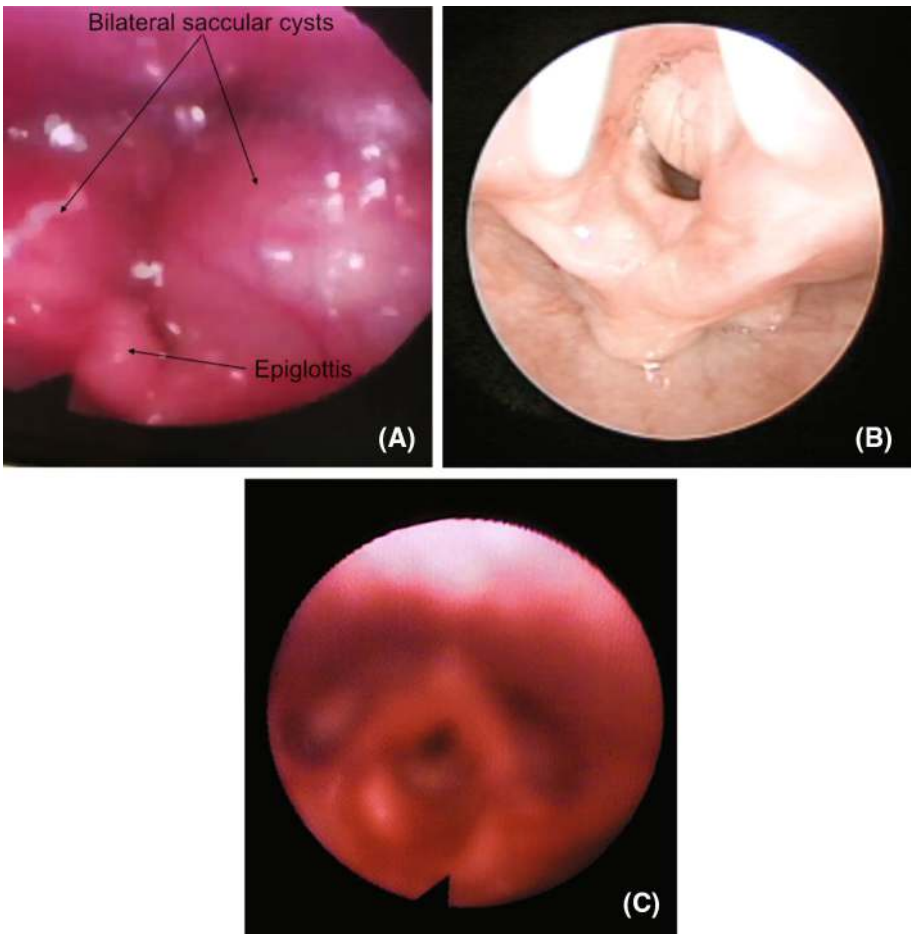

Fig. 3 Laryngoscopy showing bilateral saccular cyst with bifid epiglottis—**a** before surgery. **b** After left cyst marsupilization. **c** After bilateral cyst marsupilization. *Note* Bifid epiglottis (*black arrow*)

epiglottis using micro instruments and 4,0 vicryl rapid suture. CO_2 laser was used to cauterize the inner surface of the bifid epiglottis to create raw surface area for healing (Fig. 4). Three months postoperative the patient had a recurrent left cyst which was managed in the same way. The patient was then decannulated and was able to swallow all food consistencies.

Discussion

The laryngeal saccule is a small space that projects superiorly from the laryngeal ventricle. It is located at the level of the false vocal cords and is lined by multiple mucosal glands. Saccular disorders are secondary to abnormal dilatation of the saccule and include both laryngoceles and saccular cysts. They can be classified according to their content and location. Depending on whether it contains air, mucus or purulent material, the lesion is named either a laryngocele with a patent saccular orifice, a saccular cyst with a blocked orifice or a laryngopyocele respectively. Classification according to the cystic mass location differentiates an internal laryngocele, which is confined to the larynx and does not extend through a perforation site in the thyrohyoid membrane, from an external laryngocele, that herniates through the thyrohyoid membrane, and a combined laryngocele with dilated internal and external components [4, 8, 9].

Congenital saccular cysts are formed when the orifice of the laryngeal saccule fails to canalize into the laryngeal lumen. The saccule then fills with mucus material and dilates. Saccular disorders, however, are more commonly acquired than congenital. They may develop secondary to an obstructing lesion such as a tumor. Alternatively some studies support the association between the development of laryngoceles and increased intralaryngeal pressure [1]. This may be brought about by activities such as coughing and straining which require fixing the diaphragm in forced expiration against a closed larynx resulting in increased glottic and ventricular pressure [8, 9]. However the relationship of laryngocele to these activities may have been overstated [4].

Congenital SCs are rare, representing only 1.5 % of all laryngeal anomalies [10]. Lateral SCs protrude from the ventricular band and/or arytenoepiglottic fold while Anterior saccular cysts arise from the anterior ventricle and bulge into the laryngeal lumen between the true and false vocal cords [5, 10]. The clinical picture of saccular

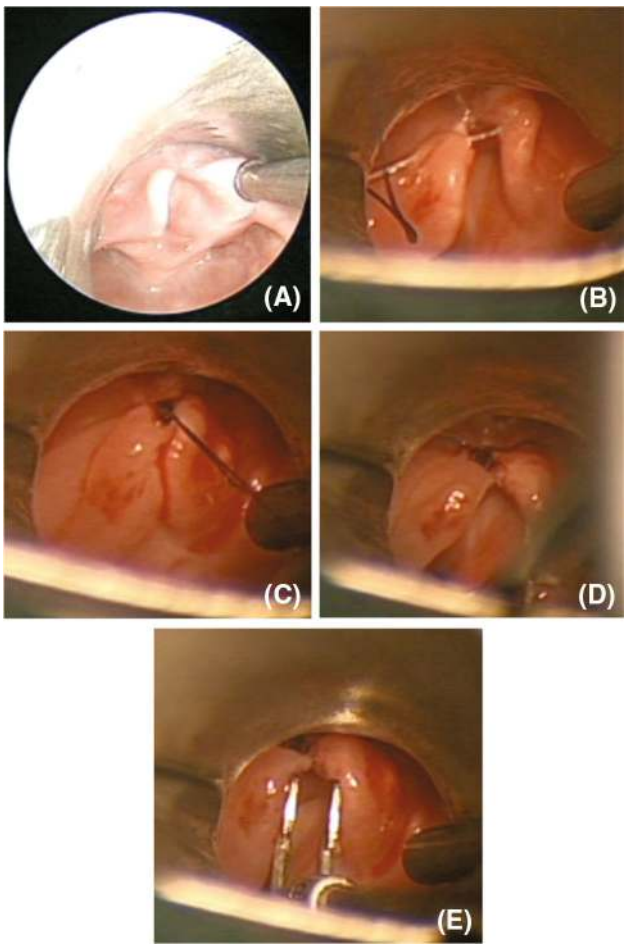

Fig. 4 Laryngoscopy showing bifid epiglottis—**a** before surgery. **b** With a needle in epiglottis. **c** After a knot approximating its two parts. **d** Sutured epiglottis. **e** Repaired epiglottis

disorders varies from asymptomatic to symptomatic according to the type, size, degree of extension and the age of the patient. If symptomatic, patients usually present with hoarseness, stridor, respiratory distress during feeding and snoring [2, 5, 10]. In rare severe cases, such as our case, saccular disorders may result in death unless immediate airway intervention occurs [11].

The diagnosis of a bifid epiglottis should prompt thorough investigations for other anomalies of PHS as laryngoscopy on 26 subjects with PHS showed that 15 had a bifid or cleft epiglottis (58 %). The most common defects are anomalies of the hands and/or feet (90 %), hypopituitarism and hypothalamic hamartomas (50 %), which is the primary feature of this syndrome [3, 12]. Other manifestations of the syndrome include, imperforate anus, renal anomalies cleft palate, cleft uvula, and laryngeal malformations [12]. The presence of a laryngocele has been reported once in association with these anomalies [1] and only one study reported a case of two saccular cysts originating from the left ventricle in association with bifid epiglottis and congenital defects including sixth digit on the left hand and an umbilical hernia [6]. We report bilateral saccular cysts. The associated anomalies include post-axial polysyndactyly of the right hand, postaxial polydactyly and extra tag on the left hand, polydactyly of the feet, microretrognathia, short neck and hypospadias. MRI of the brain was unremarkable with no evidence of hypothalamic hamartomas.

Management of saccular cysts remains controversial. It includes maintenance of airway and cyst removal. Maintenance of oxygenation can be achieved by cricothyrotomy, intubation, tracheotomy or even cardio-pulmonary bypass may be required in extreme situations. The treatment options for cyst removal are either external or internal approaches. The external approach is usually reserved for recurrent cases. The internal approach is via microlaryngoscopy and is the most common choice including endoscopic needle aspiration, marsupialization and ventriculotomy [2, 10, 13]. Endoscopic marsupialization of the cyst with application of CO_2 laser proved to be safe and effective [10, 13]. Endoscopic radiofrequency ablation is a new technique reported in the literature that may improve surgical precision and better healing [7]. Another reported technique involved using a 30° angled telescope with a flexible laser fiber system that provided excellent visualization of the saccular cysts [14].

Management of bifid epiglottis is mostly conservative with excellent results. It includes supporting the airway as needed and correcting the associated anomalies. The patient will need follow up and observation for the presence of a lax epiglottis, which may prolapse into the laryngeal inlet and lead to aspiration, chocking, stridor and airway obstruction. Interventions used to manage lax epiglottis after follow up includes supraglottoplasty, epiglottectomy that yielded good results. Attempts were reported to repair the defect by trimming the edges of the epiglottis with a CO_2 laser and then suturing the two cartilaginous halves together but eventually epiglottectomy was done and resulted in a good outcome after the patient presented with symptoms of airway obstruction [4, 15–17].

For those cases of saccular cysts reported in the literature associated with bifid epiglottis, management included maintenance of the airway via treacheostomy and subsequent removal of the cysts using the Jako-laryngoscope with the surgical microscope. No surgical intervention was done for the bifid epiglottis [6].

Conclusion

Congenital laryngeal anomalies such as saccular cysts are rare but one needs to consider them in cases of neonatal respiratory distress. In the presence of a bifid epiglottis

thorough assessment for associated anomalies and syndromes should be performed. Surgical management of bifid epiglottis is reserved for those cases with symptoms of aspiration or sleep apnea.

Compliance with Ethical Standards

Conflict of interest Rawan Arif, Talal Al-Khatib, Razan Daghistani and Maher Shalbi have declare that they have no conflict of interest.

Human and Animal Rights This article does not contain any studies with human participants performed by any of the authors and does not contain any studies with animals performed by any of the authors.

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