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Case Report

Management of congenital cartilaginous sleeve trachea in children

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ABSTRACT

Aims: Children with congenital tracheal cartilaginous sleeve may present to otolaryngology services with airway problems. We wish to describe our overall management in a series of four children with this very rare anomaly.
Methods: Retrospective case note review of children diagnosed with congenital tracheal cartilaginous sleeve presenting to our department between 2006 and 2014.
Results: Four patients were seen. One had Opitz G syndrome, two had Pfeiffers syndrome and one had no associated anomalies. Two children were successfully managed with laryngeal reconstruction using an anterior costal cartilage graft, while the third and fourth required a short period of tracheostomy only. All four are well and currently asymptomatic from an airway point of view.
Conclusion: Congenital tracheal cartilaginous sleeve is a very rare and potentially challenging problem. Otolaryngologists should be aware that it can occur in children with syndromes other than craniosynostosis (and indeed, those with no syndrome) and that it can be successfully treated using established airway management techniques.
Ethical approval: Registered with Clinical Governance Committee.

1. Introduction

Congenital tracheal cartilaginous sleeve is a very rare congenital malformation in which cartilaginous rings are fused anteriorly to form an uninterrupted cartilaginous sleeve. The trachea has an abnormally smooth appearance without normal ridges or visible tracheal rings, but the trachealis muscle is still visible posteriorly, as the cartilaginaous sleeve retains the c-shape of normal tracheal rings. The sleeve can be isolated to one or two tracheal rings, usually the first and second ring, but may extend down a variable distance into the trachea or even into the bronchi [1–3].

The lesion is very much distinct from the congenital complete rings anomaly, which usually affects the distal trachea near the carina. This has distinct rings, which are not fused to each other, and the trachealis muscle is not visible [see Fig. A1].

Congenital tracheal cartilaginous sleeve has been most commonly reported alongside syndromic craniosynostosis (Crouzon's and Pfeiffer's syndromes) and occasionally with Goldenhar's syndrome. In craniosynostosis it is thought that abnormal fibroblast growth is associated with sleeve trachea, in particular fibroblast growth factors 10 and 2 [4,5]. The embryological explanation remains unclear [6]. Presentation is with stridor and respiratory difficulty. Tracheostomy may be required in the neonatal period [7], and is generally associated with decreased morbidity levels [2,8].

Few cases have been described in the literature, and many otolaryngologists will have limited experience of the condition. Previous studies have indicated that management of infection and secretions, appropriate tracheostomy tubes and endoscopy are vital in looking after children with congenital tracheal cartilaginous sleeve [9]. We therefore report on our airway management in children with this anomaly.

2. Methods

A retrospective case note review of all children diagnosed with congenital tracheal cartilaginous sleeve presenting to the Royal Hospital for Sick Children, Glasgow between 2006 and 2014. Cases were identified from the departmental database and personal records of the surgeons in the department. All otolaryngological symptoms, signs, investigations and treatments were recorded and analysed.

3. Results

3.1. Case 1

This 2-month old female presented to our services with, dysmorphic facial features, cranial vault expansion, hydrocephalus and a raised intra-cranial pressure consistent with a diagnosis of

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syndromic craniosynostosis. In addition, she was also suffering from upper airway obstruction with poor feeding, intercostal indrawing, stridor, sleep apnoea and obvious respiratory distress. She had a genetic verification of Pfeffier's syndrome. No cleft lip or palate was present.

Computed-tomography showed mid-face retrusion, a degree of forehead brachycephaly and supraorbital retrusion. Overnight and daytime-nap pulse oximetry studies were performed, demonstrating periods of oxygen desaturations. Supplemental oxygen therapy was commenced. Awake transnasal fibreoptic laryngoscopy showed copious clear secretions in the nasal cavity and pharynx along with airway obstruction at the pharyngeal level. A nasopharyngeal airway was trialled and this improved the stridor, indrawing and poor feeding to some extent. Residual oxygen desaturations and feeding difficulties however, led to the decision to perform a tracheostomy at 2 months. At tracheostomy, the microlaryngobronchoscopy showed that the 2nd, 3rd and 4th tracheal cartilages were fused anteriorly to form a complete congenital tracheal cartilaginous sleeve.

Once the tracheostomy was in place (size 3.5, flexed end) the child was weaned off oxygen and began eating and drinking well. A ventriculo-peritoneal shunt was inserted for hydrocephalus as an emergency procedure, leading to a short-lived period of increased respiratory secretions, apnoeic episodes and oxygen desaturations. No further complications have been experienced, including choking, gagging or respiratory infections, and a mid-face advancement may or may not facilitate decannulation.

3.2. Case 2

This girl was referred to us at the age of 10 years, having had a tracheostomy performed in infancy at another hospital for breathing difficulties. She had been diagnosed with Opitz G syndrome, was wearing hearing aids for a sensorineural hearing impairment and also had undergone a cleft palate repair.

She was known to have a very difficult view at laryngoscopy, and airway examination in her referral to hospital had been incomplete. A computed-tomography scan had therefore been performed to provide more detailed information on airway anatomy. The scan suggested a significant stenosis of the upper trachea above the tracheostomy. Microlaryngobronchoscopy under general anaesthetic confirmed a very difficult view at laryngoscopy preventing confident diagnosis of the nature of the airway anomaly, although it was clear that there was a degree of stenosis of the upper trachea, a bifid, short epiglottis, shortened vocal cords and oedema over the arytenoids as a result of gastro-oesophageal reflux disease. It was felt that a laryngeal reconstruction would be required to achieve tracheostomy decannulation but this was deferred for a year as the child was being considered for pharyngoplasty surgery for velopharyngeal incompetence.

At 11 years of age she underwent laryngotracheal reconstruction with an anterior costal cartilage graft as a single stage procedure with removal of the tracheostomy, followed by a week of post-operative intubation. At surgery the nature of the upper tracheal stenosis became apparent: there was a congenital tracheal cartilaginous sleeve with fusion of the 2nd, 3rd and 4th tracheal cartilages anteriorly but preservation of the trachealis muscle posteriorly. The procedure went well. Within 1 week the graft was completely musossalised and both the subglottis and trachea were patent with no residual stenosis at final endoscopy at 2 weeks. Upon review 2 years later the patient was doing well with no exercise restrictions, no stridor and a strong voice.

3.3. Case 3

This 4-month old male presented at a nearby district hospital with stridor, which had been present since birth, and poor feeding. He was born at full term and had never been intubated. He was given nebulised epinephrine and oral dexamethasone on presentation, but his stridor and respiratory distress persisted and so he was referred to our department.

On examination he had biphasic stridor at rest with cough, tracheal tug and sternal recession. Microlaryngobronchoscopy showed a 70% stenosis in the subglottis that was thought to be congenital subglottic stenosis. There was also a mild degree of tracheomalacia. He was given anti-reflux medication and observed clinically in the hope that his symptoms would improve with growth. Over the subsequent 12 months it became clear that his breathing difficulties were getting worse rather than better however, with significant exercise limitation, recurrent episodes of croup and increasing stridor at rest. Therefore at 18 months of age he underwent single stage laryngeal reconstruction with an anterior costal cartilage graft.

It was apparent at open surgery that the trachea was very unusual in appearance with a congenital cartilaginous sleeve trachea anomaly with fusion of the 1st, 2nd and 3rd tracheal cartilages and a preserved trachealis muscle posteriorly [see Fig. A2]. He spent one week intubated after surgery, at which point endosocopy showed a normal-calibre airway. He was asymptomatic at review with no exercise limitation or stridor and a strong voice.

3.4. Case 4

This girl presented to our department at 4 months old. She was transferred from another hospital after two admissions with ongoing respiratory issues, including respiratory arrest, apnoeas and obstructive airway especially at night. She presented with chronic airway obstruction in relation to Pfeffier syndrome Type 2 and associated craniosynostosis, cervical spinal fusion, hydrocephalus with a VP shunt, bilateral hearing aids, noisy breathing and obstructive sleep apnoea. She has marked frontal bossing, an abnormal shaped skull with shortening in the back to front dimension, proptosis of the eyes and sacral eversion producing a prominence at the nasal cleft [see Figs. A2 and A3].

She underwent microlaryngobronchoscopy and tracheostomy in our department, which was successful however she complicated post-operatively with a likely pneumonia, which was given antibiotic treatment. Microlaryngobronchoscopy showed a complete congenital tracheal cartilaginous sleeve [see Fig. A1].

She then underwent cranial vault expansion, mid-face and forehead vault destractions to relieve upper airway obstruction between 6 and 10 months and was decannulated at 11 months after a further microlaryngobronchoscopy, which showed no structural deformity. Since she has been using 0.5l of 99% O_2 at night, indicated by a sleep study at that time, which showed 8 desaturations per hour.

At 16 months she underwent elective tonsillectomy, adenoidectomy and surgical closure of her tracheostomy. The operation went well with no complications. Recovery from anaesthetic was slow however and this raised questions as to whether she was experiencing central apnoeic episodes. Resulting sleep studies showed desaturations and raised CO_2 levels overnight. She was also experiencing a lot of secretions, which improved with saline nasal drops. Her sleep study then improved and it was felt that her desaturations were secondary to obstruction from the secretions. She was then fit for discharge, with a view to review sleep studies again in future.

4. Discussion

Congenital tracheal cartilaginous sleeve is sufficiently rare that many otolaryngologists will have never encountered it. In a child with syndromic craniosynostosis, airway obstruction may be blamed on the obvious mid-face hypoplasia but the otolaryngologist should be aware that there may be other causes, with congenital