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CLINICAL RECORD

Difficult airway in a child with severe dystonia

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Abstract

Objective: To describe the management of a 15-year-old girl with repeated life-threatening complications of her tracheostomy secondary to muscle dystonia and thoracolumbocervical lordosis.

Method: This paper reports a retrospective case review.

Results: Regular microlaryngoscopy and bronchoscopy, treatment with systemic steroids and a soft tracheostomy tube, in addition to better control of the dystonia, resulted in control of the patient's airway. This minimised tracheal inflammation and granulation tissue formation.

Conclusion: The need for a tracheostomy in patients with thoracolumbocervical lordosis and severe dystonia should be considered only after all other options of airway management have been explored. Every attempt should be made to minimise tracheal trauma caused by excessive movement of a tracheostomy tube.

Key words: Tracheostomy; Child; Dystonia; Tracheomalacia; Granulation Tissue

Introduction

Complications of tracheostomy are varied and include obstruction of the tube by granulation tissue. In isolation, these can be managed using a variety of techniques. In combination with tracheomalacia caused by abnormal posture and repeated dystonic motion, these complications can be challenging to treat and may compromise the airway.

We present the case of a 15-year-old girl with thoracolumbocervical lordosis. The patient had a tracheostomy in situ and suffered from severe dystonic contractions, which presented a challenging case for airway management. The patient's posture and abnormal dystonic movements resulted in tracheomalacia and severe tracheal granulations. This caused repeated life-threatening respiratory distress associated with occlusion of the tracheostomy tube.

Case report

A 15-year-old girl with cerebral palsy was admitted with a lower respiratory tract infection. Due to the respiratory compromise, she was intubated and ventilated. Extubation, which was attempted three times, was not possible during recovery from the infection because of upper airway obstruction. A tracheostomy was performed in order to facilitate extubation. This was soon followed by periods of respiratory compromise. These were triggered by dystonic contractions of the upper body wherein the thoracic and cervical spine regions were hyperextended. The use of a bag valve mask was adequate for resuscitation.

Rigid and flexible endoscopic examination of the airway revealed extensive tracheal granulations and distal tracheal tracheomalacia (Figure 1). The airway was assessed with the thoraco-cervical spine in both a neutral position and in the hyperextended position which mimicked the dystonic events. This revealed that the tracheostomy tube was occluded during hyperextension as a result of granulations on the anterior tracheal wall.

The respiratory arrests were prevented by attaining better control of the dystonic events. This involved the use of combination treatment comprising titrated diazepam, anticholinergics and carbamazepine. The tracheal granulations were treated with steroids in the form of topical dexamethasone drops (0.05 per cent) applied twice daily to the stoma and trickled into the trachea, and oral prednisolone (1 mg/kg/day). Co-amoxiclav was also administered to treat the infective component of the granulations. In addition, a Tracoe Vario tracheostomy tube Kapitex (Wetherby, UK) was used, which is softer and more flexible than other tubes, making insertion less traumatic.

Discussion

Patients with severe motor and intellectual disabilities associated with thoracolumbocervical lordosis and dystonic contractions can present a challenging case for surgical airway management. Many of these patients have narrowing of the mediastinum caused by scoliosis and lordosis, which is exacerbated by dystonic contractions. These abnormal hyperextended positions and repetitive abnormal movements may lead to recurrent serious and life-threatening complications should a tracheostomy be in situ. Such complications are clinically challenging due to persistent tracheal irritation.[1]

A surgical tracheostomy should only be performed in patients with severe motor and intellectual disabilities as a final option, after all other avenues of airway management have been explored. Despite best efforts, these patients commonly undergo tracheostomy following emergency endotracheal intubation for acute respiratory failure.[2] Several complications have been observed, such as tracheal granulations, tracheomalacia and tracheo-innominate artery fistula.

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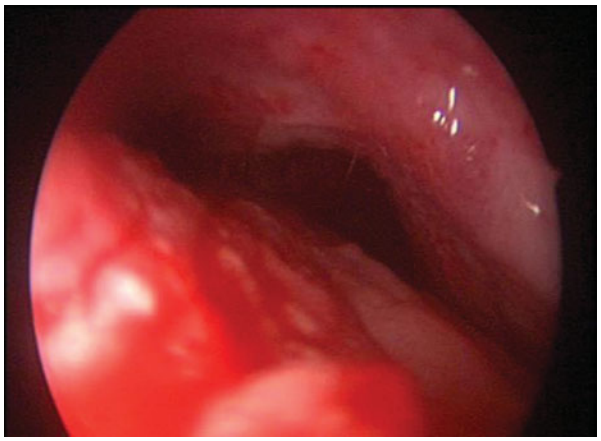

FIG. 1
Intra-operative endoscopic view of the trachea showing tracheomalacia and tracheal granulations.

Self-made and made-to-order long tracheostomy tubes have been described for the management of tracheomalacia and tracheal granulations.

- **Tracheostomy in the paediatric population is uncommon and complications include tracheomalacia and granulations**
- **Tracheostomy in a dystonic patient can additionally lead to occlusion due to abnormal posture and movement**
- **These patients should receive a tracheostomy only after all other options of airway management have been explored**

Granulations associated with the use of tracheal tubes have been reported in patients with severe motor and intellectual disabilities.[3] They tend to occur on the anterior wall of the trachea and are exacerbated by torticollis, cervical lordosis and thoracic deformity. Treatments have included changing the tracheostomy to an adjustable flange tube, using a laser, and the use of mitomycin C in relapsing cases. It is imperative that the tracheostomy tube is made of a soft material, and that it has a suitable length and angle in order to limit tracheal irritation and granulation formation.[4]

In summary, the management of tracheostomy in patients with severe motor and intellectual disabilities can be challenging and life-threatening. The abnormal posture predisposes the patient to tracheomalacia, and the dystonic events encourage granulation tissue formation. This combination of conditions can have catastrophic sequelae if the tracheostomy tube becomes blocked by granulations on the tracheal wall. This cohort of patients should receive a tracheostomy only after all other options of airway management have been explored.

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