CASE REPORT

Painful torticollis following adenotonsillectomy: a cardinal sign of atlantoaxial subluxation

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SUMMARY

An 11-year-old boy with a history of autism spectrum disorder attended the emergency department with his mother 8 days after an adenotonsillectomy reporting postoperative bleeding. Detailed physical examination revealed no active bleeding, but a rigid neck posture was noted. A head and neck CT scan demonstrated unilateral rotatory atlantoaxial subluxation and possible damage to the anterior spinal ligament. He was reviewed by neurosurgeons who performed manipulation under anaesthetic and successfully realigned the occipital cervical tract. Non-traumatic atlantoaxial subluxation (Grisel's syndrome) is a rare but serious complication of routine ear, nose and throat (ENT) procedures. An awareness of this complication among paediatricians, otolaryngologists and emergency physicians, and a high index of suspicion in any patient presenting with torticollis following ENT surgery is essential in preventing significant neurological morbidity.

BACKGROUND

Postoperative pain is a common complaint following routine ear, nose and throat (ENT) procedures such as adenotonsillectomy, and in most cases is successfully managed with simple analgesia and rest. While pain can limit an individual's range of movement, any significant restriction in range of neck movement or fixed neck posture should raise suspicion of an underlying atlantoaxial injury, a rare but extremely important and often overlooked complication of adenotonsillectomy. The case we describe is of particular importance to those working in general practice, emergency medicine, paediatrics and ENT, who do not infrequently encounter patients complaining of pain following this common procedure.

CASE PRESENTATION

An 11-year-old boy presented to the emergency department 8 days following an uneventful adenotonsillectomy with postoperative bleeding. The adenoidectomy had been performed with monopolar suction diathermy, and the tonsillectomy with bipolar diathermy. His mother reported that he had coughed up a small quantity of blood in the early hours of the morning. He had a medical history of autism spectrum disorder and was otherwise fit and well. On presentation, he was haemodynamically stable, and on examination of the oropharynx, there was a small quantity of blood staining in the right tonsillar fossa but no active

bleeding. On detailed physical examination, markedly abnormal head posture was noted, including an inability to bring his head into midline, with no associated neurological deficits ([video 1](#)). His mother reported that from day 1 postoperatively, she had been concerned about a stiff neck, and had previously taken him to a walk-in centre where he was assessed and discharged with simple analgesia.

INVESTIGATIONS

A diagnosis of non-traumatic atlantoaxial subluxation was suspected and a CT scan of the neck performed, which demonstrated unilateral rotatory atlantoaxial subluxation, with more than 75% of the right lateral mass of C1 displaced anteriorly and likely caused damage to the anterior spinal ligament ([figure 1](#)).

TREATMENT

The patient was initially managed with a soft collar, simple analgesia and muscle relaxants and an urgent neurosurgical review was sought. In addition, intravenous antibiotics and hydrogen peroxide gargles

Video 1 Video of the patient taken on admission demonstrating a neck posture in fixed lateral rotation typical of atlantoaxial subluxation.

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Figure 1 CT of the cervical spine. These axial slices demonstrate greater than 75% anterior subluxation and rotation of the right lateral mass of C1, and an associated lateral shift of the dens to the left.

were prescribed in the context of a post tonsillectomy bleed. Difficulty was noted in administering oral medications including muscle relaxants due to the patient's autism spectrum disorder, and on neurosurgical review, it was determined that conservative management was unlikely to bring about resolution. He underwent manipulation under anaesthesia of C1 and C2 the following day. The procedure used an established three step manual-reduction technique, comprising (1) lateral mobilisation of the cervical spine in the coronal plane, (2) lateral neck flexion to both sides and (3) rotation in the longitudinal axis.¹² The intervention resulted in good alignment of cervical occipital tract and he was discharged the same day with a hard cervical collar.

OUTCOME AND FOLLOW-UP

On 6-week outpatient review, the collar was removed and he was noted to have retained good cervical alignment and symmetrical

but reduced neck movements due to discomfort. A subsequent MRI has demonstrated maintained good alignment of the cervical occipital tract and he continues to receive follow-up with neurosurgery.

DISCUSSION

Non-traumatic atlantoaxial subluxation (Grisel's syndrome) is a rare but important condition, with only 105 cases documented in the literature between 1930 and 2016.³ In the ENT context, it is most frequently seen as a complication following adenotonsillectomy.⁴ It can also occur as a sequelae to upper respiratory tract infections, and while its aetiology remains uncertain, a commonly accepted hypothesis is that spread of infection from the posterior pharynx to the periodontoid venous plexus leads to pathological laxity of atlantoaxial ligaments and greater joint instability.^{3,5} Pre-existing atlantoaxial ligamentous laxity, as seen in children and patients with Down's syndrome, is an additional risk factor for the development of the condition.^{3,6} The use of monopolar suction diathermy in adenoidectomy, as was used in this case, is also a recognised risk factor, with a retrospective review of 1431 adenoidectomies observing a rise in the rate of Grisel's syndrome from 0% to 0.4% following introduction of the technique in one centre.⁷ The risks associated with monopolar suction electrocautery must be weighed against its demonstrated benefits of reduced rates of intraoperative haemorrhage and reduced operative time.⁸

While no body-approved guidelines exist for the diagnosis and management of Grisel's syndrome, in the reported literature, a high index of suspicion in addition to CT or MRI have invariably been necessary in confirming the diagnosis and severity of subluxation/rotation.⁴ Severity of injury is graded using the Fielding classification (types I to IV).⁹ Type I is a fixed but purely rotational deformity, with a normal atlantodental interval (2 mm). In types II and III, there is fixed rotation and anterior dislocation of the atlas with an increased atlantodental interval of 3–5 mm and >5 mm, respectively. Type IV describes a rotational deformity with posterior dislocation of the atlas. A majority of cases are Fielding types I and II, and are successfully treated with intravenous antibiotics, simple analgesia, muscle relaxants and immobilisation. Type III subluxations, as in the case we describe, are often refractory to medical therapy, and require closed reduction and immobilisation, while in type IV, internal fixation or C1-2 fusion are often indicated.^{2,9} This algorithm serves as a guide to management but must be complemented by an individualised approach, for example, with more aggressive management considered in type I and II subluxations where there are associated neurological deficits, or that fail less invasive measures.⁴

In the case we have presented, it is likely that limited oral intake postoperatively had an important role in the development of localised infection, which in turn contributed to a self-limiting postoperative haemorrhage and atlantoaxial subluxation. Diminished oral intake was due to pain and poor compliance with analgesia, the latter of which in this instance was linked to underlying autism spectrum disorder. It is worth highlighting that had the post-tonsillectomy haemorrhage required surgical management, achieving the requisite neck extension would have been precarious, given the underlying atlantoaxial injury. While this case depicts a delayed diagnosis of Grisel's, it is unclear whether earlier presentation and recognition would have resulted in successful non-operative management.

An awareness of Grisel's syndrome and a corresponding high index of suspicion are essential among paediatricians,

otolaryngologists and emergency physicians when assessing patients with painful torticollis following ENT surgery. Systematic review of the literature found this to be the first documented case of Grisel's syndrome occurring in an individual with autism spectrum disorder, and as our case demonstrates, early neurosurgical intervention may be of benefit when administration of analgesia and muscle relaxants are poorly tolerated.

Learning points

- ▶ Atlantoaxial subluxation is a rare but important complication of ear, nose and throat surgery that otolaryngologists, paediatricians and emergency medicine physicians must be aware of to ensure timely diagnosis and prevent morbidity.
- ▶ In patients with painful torticollis following adenotonsillectomy, atlantoaxial subluxation should be the presumptive diagnosis pending further investigation.
- ▶ Urgent CT/MRI of the neck is crucial in confirming the diagnosis.
- ▶ Immediate management includes cervical collar immobilisation, simple analgesia, muscle relaxants and urgent neurosurgical consultation.
- ▶ Special attention should be afforded to individuals with behavioural disorders, who, if unable to tolerate conservative treatment options, may benefit from early neurosurgical intervention.

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