CASE REPORT

Torticollis following tympanoplasty: an index case with lessons in encountering the unexpected

Krishan Ramdoo,¹ Andy Hall,¹ Panagiotis A Dimitriadis,² Arvind Singh¹

¹Department of Ear, Nose and Throat Surgery, Northwick Park Hospital, London, UK

²Department of Ear, Nose and Throat Surgery, Luton and Dunstable Hospital, Luton, UK

Correspondence to
Krishan Ramdoo,
kramdoo@hotmail.com

Accepted 18 February 2014

SUMMARY

The term torticollis is derived from the Latin words 'tortis' for twisted and 'collum' for neck. Typical features include sudden onset of severe unilateral pain, with restriction of movements and deviation of the neck to that side. Although torticollis is a well recognised abnormality, there is minimal literature identifying it as a sole long-standing complication in otological surgery. We present the case of a 15-year-old boy who developed permanent torticollis following a routine tympanoplasty. A CT of the neck revealed an atlantoaxial rotatory fixation with cross fusion between C1 and C2. This case highlights a rare cause of torticollis resulting from a routine otological procedure. It also differentiates between Grisel's syndrome, which is characterised by atlantoaxial subluxation as opposed to fusion. The role of the multidisciplinary team is imperative and in particular the need for urgent discussion with the neurosurgical team to ensure prompt assessment and management of the patient takes place.

BACKGROUND

The term torticollis is derived from the Latin words 'tortis' for twisted and 'collum' for neck. This definition is adopted by the National Institute for Health and Care Excellence (NICE) that states that acute torticollis is secondary to minor local musculoskeletal irritation causing pain and spasm in neck muscles. Typical features include sudden onset of severe unilateral pain, with restriction of movements and deviation of the neck to that side. It is most commonly congenital or postnatally acquired.^{1 2}

Although torticollis is a well recognised abnormality, there is minimal literature identifying it as a sole long-standing complication in otological surgery. We present the case of a 15-year-old boy who developed permanent torticollis following a routine tympanoplasty.

CASE PRESENTATION

A boy with severe learning difficulties had been presenting frequently to the ear, nose and throat (ENT) department with bilateral wax since the age of 5 years. He had an extensive medical history consisting of previous craniofacial surgery for sagittal craniosynostosis and leg-length discrepancy with talipes. He had grommet insertion when he was a child and had some mixed conductive and sensorineural hearing loss. However, there was no formal diagnosis of any developmental syndrome at this stage. There was no significant family history of hearing loss or any other systemic disease.

Despite regular review in the outpatient clinic and the examination of the ears under anaesthetic, at the age of 13, he underwent an uncomplicated left tympanoplasty with cartilage reconstruction for a persistent perforation of the tympanic membrane from recurrent infections. A postauricular approach was used in the standard fashion and the procedure was successful. Anaesthetically, there were no difficulties and there was no unusual rotation of the head.

Immediately postoperatively, however, a right-sided torticollis developed. It was initially deemed due to muscle spasm and thought likely to settle over time and he was prescribed regular analgesia and muscle relaxants to help relieve the symptoms.

Six months postprocedure, the torticollis had not settled despite receiving physiotherapy and a botulinum toxin injection into the sternocleidomastoid muscle. Although the pain had eased, the torticollis posture had persisted and his overall neck and alignment had not returned to its normal state. After liaising with the paediatric neurosurgical team, a CT scan and an MRI were organised.

INVESTIGATIONS

The CT identified a severe atlantoaxial rotatory fixation with cross fusion between C1 and C2, the possibility of an undiagnosed congenital fusion between C2 and C3 (Klippel-Feil anomaly) and thoracic scoliosis (compensatory). The MRI showed that there was no involvement of the spinal cord and there was no obvious compression around the cervicomedullary junction. The fact that the rotatory deformity was fixed made manipulation or traction an unlikely option at this stage.

DIFFERENTIAL DIAGNOSIS

Torticollis, muscle spasm and Grisel's syndrome.

TREATMENT

A multidisciplinary approach was used throughout among the ENT team, paediatricians, neurosurgeons and physiotherapists. However, the cross-fusion of C1 and C2 shown on the CT meant that surgical intervention would be technically difficult and of high risk with only a moderate chance of improvement. A decision was made by the paediatric neurosurgical team not to proceed with surgery on the cervical spine and a referral was made to the spinal orthopaedic team to assess the possibility of correction of the thoracic scoliosis.

CrossMark

To cite: Ramdoo K, Hall A, Dimitriadis PA, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2013-201539

Figure 1 Serial axial cuts of CT of the neck demonstrating the atlantoaxial rotatory fixation.

OUTCOME AND FOLLOW-UP

This patient is being seen on a regular basis by the orthopaedic team. A follow-up MRI is being organised to see how he progresses.

DISCUSSION

This case highlights a rare cause of torticollis resulting from a routine otological procedure. A particular 'learning point' is to highlight this as a potential complication of otological surgery where a prolonged period of head rotation may be required.

Muscle spasm can occur following the otological surgery; however, advice from the neurosurgical team would help to identify the need for urgent imaging, intervention or assessment. In a child where muscular aetiology for torticollis has been excluded, other neurological abnormalities or Klippel-Feil anomalies may be the underlying causes.³

The management should begin at the start of the patient's journey when consenting the patient for theatre. Although it poses a low risk, it should be mentioned to the parents the

Figure 2 Thoracolumbar scoliosis.

Figure 3 Arrows pointing to atlantoaxial fixation between C1 and C2 (sagittal view). Red vertical line shows congenital cross-fusion between C2 and C3 (Klippel-Feil anomaly).

possibility of atlantoaxial subluxation or fixation following an otology procedure.⁴ There is a school of thought that excessive passive rotation and/or hyperextension of the head as well as

transferring the patient without a roll board could lead to increased incidence of atlantoaxial subluxation.⁴ This needs to be avoided in all cases to prevent the possibility of fixation too.

Figure 4 Fielding-Hawkins classification of C1–C2 rotary subluxation. Type I: rotatory fixation without anterior displacement of atlas (≤ 3 mm). Type II: rotatory fixation with anterior displacement of atlas of 3–5 mm. Type III: rotatory fixation with anterior displacement of atlas of >5 mm. Type IV: rotatory fixation with posterior displacement of atlas.

Minimising delays in discussion with specialist teams will also help to prevent fusion from occurring which hinders the possibility of less invasive intervention such as traction being employed. The difficulty would be to ensure that this would not be overdiagnosed and prompt clinical assessment would need to be the basis for referral.

Grisel's syndrome is a phenomenon that has been previously identified as a complication of several otolaryngology pathologies such as with pharyngitis, adenotonsillitis, tonsillar abscess and otitis media due to inflammatory laxity of the upper cervical ligaments.⁴ It can commonly present with torticollis; however, its true definition is atlantoaxial subluxation not associated with trauma or bone disease.⁴ If left untreated, it may lead to degenerative disorders of the cervical spine or even with neurological malfunction.⁵ In the case we present it is atlantoaxial rotatory fixation (figure 1), which was the defining feature distinguishing it from Grisel's syndrome; also none of the above-mentioned complications occurred and having developed some thoracolumbar scoliosis, it may have hindered recovery (figure 2). The difficulty in this case is in identifying whether the prolonged torticollis led to atlantoaxial fixation or whether there was an element of subluxation initially which developed fixation. Having a CT of the neck at an earlier date rather than 6 months later may have enabled us to define this. The reason for the delay in our case was that it was thought the torticollis would settle and it was actively being treated with regular analgesia and muscle relaxants. An incidental finding on the CT was the presence of congenital fusion of C2 and C3 (Klippel-Feil anomaly) (figure 3). This is characterised by bony abnormalities, usually fusion of the cervical vertebral bodies. It may present clinically with short neck, reduced cervical range of movement and a low posterior hairline.⁶ In a study of 288 children with torticollis, 53 had a non-muscular aetiology for torticollis and 30% of them had Klippel-Feil anomaly.³ Although the patient did not exhibit all these characteristics, it should still be considered as a possible cause.

Despite not having true clarity on whether subluxation may have occurred prior to fixation, it should be said that the management would be the same. A CT and three-dimensional reconstruction can be pivotal at an early stage and if true subluxation was present, then different management approaches would be used depending on the Fielding-Hawkins classification of C1 and C2 subluxation³ (figure 4). Types 1 and 2 would be treated with antibiotics and immobilisation in a cervical collar. Only in some cases, cervical traction would be required. For types 3 and

4, bed rest and cervical traction would be required. If there is a failure in these treatment methods, a surgical opinion would be sought, as in our case. This management, however, should still be discussed with the neurosurgical team as soon as the symptom is noticed.

Learning points

- ▶ Torticollis is a recognised complication following otological surgery. Urgent neurosurgical review should be requested in all cases to ensure prompt assessment and management.
- ▶ Klippel-Feil anomaly or other neurological abnormalities may be the underlying pathologies in the absence of muscular causes of torticollis.
- ▶ Assessing and managing the paediatric population is challenging and even more so if the problem is not a recognised one.
- ▶ Early discussion in a multidisciplinary forum would allow clear guidance and advice about management; the patient and parents should be kept informed at all times.

Contributors PAD was involved in study design, acquisition and analysis of data, literature review; submission process and final approval of the version to be published. KR was involved in manuscript writing, study design, literature review and final approval of the version to be published. AH was involved in conception of study, obtained consent from patient, interpretation of data and final approval of the version to be published. AS was involved in study design, drafting of the article and final approval of the version to be published.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Tomczak KK, Rosman NP. Torticollis. *J Child Neural* 2013;28:365–78.
- 2 NICE. Neck pain – acute torticollis. <http://cks.nice.org.uk/neck-pain-acute-torticollis#topicsummary> (accessed 23 Jan 2014).
- 3 Ballock RT, Song KM. The prevalence of nonmuscular causes of torticollis in children. *J Pediatr Orthop* 1996;16:500–4.
- 4 Bocciolini C, Dall'Olio D, Cunsolo E, et al. Grisel's syndrome: a rare complication following adenoidectomy. *Acta Otorhinolaryngol Ital* 2005;25:245–9.
- 5 Hirth K, Welkoborsky HJ. [Grisel's syndrome following ENT-surgery: report of two cases]. *Laryngorhinootologie* 2003;82:794–8.
- 6 Balci S, Oguz KK, Firat MM, et al. Cervical diastematomyelia in cervico-oculo-acoustic (Wildervanck) syndrome: MRI findings. *Clin Dysmorphol* 2002;11:125–8.

Copyright 2014 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow