



REVIEW ARTICLE

Grisel's syndrome in otolaryngology: A systematic review

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Received 21 May 2007; received in revised form 5 July 2007; accepted 6 July 2007
Available online 13 August 2007

KEYWORDS

Grisel's syndrome;
Spontaneous non-
traumatic atlantoaxial
subluxation;
Head;
Neck;
Ear;
Nose;
Throat

Summary

Aims: to assess etiology, treatment and outcome of Grisel's syndrome.

Methods: A Medline search was performed using the terms Grisel's syndrome, spontaneous atlantoaxial subluxation, head, neck, ear, nose and throat. A systematic review of the literature was performed. Case series of both adult and pediatric cases were included. Only papers focusing on true non-traumatic atlantoaxial subluxation were included.

Results: Seventy-one papers have been published from 1950 to 2006. Forty-eight of these fulfilled our inclusion criteria, totaling 103 patients for review. The main causes of Grisel's syndrome were infection (48%) and post-adenotonsillectomy (31%). Less common causes included other postoperative cases such as pharyngoplasty and ear operations. Neurosurgical consultation was paramount in all cases. In the majority of cases conservative management in the form of bedrest, antibiotics, muscle relaxants, traction and collar was effective; in a few cases only surgery in the form of arthrodesis was deemed necessary. Morbidity was significant in those cases where diagnosis was delayed, with the most devastating consequence a permanent neurological deficit in one case.

Conclusions: Grisel's syndrome is a rare but dangerous complication that can go unnoticed in its early phase and can be a major cause of morbidity and mortality following infection or head and neck procedures/interventions. Early recognition of any cervical complication following routine otolaryngological operations together with early neurosurgical consultation is mandatory to prevent devastating consequences.

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Contents

1. Introduction	1824
2. Methods	1824
3. Results	1824
4. Discussion	1824
5. Conclusion	1826
References	1826

1. Introduction

Cervical complications following head and neck procedures are uncommon but, when they do occur, they are associated with significant morbidity, especially when not promptly diagnosed. Non-traumatic atlantoaxial subluxation (NAAS) was first described by Charles Bell in a patient with syphilis, pharyngitis and lethal spinal compression [1]. In 1951, Grisel described two cases of pharyngitis and atlantoaxial subluxation [1].

Grisel's syndrome refers only to non-traumatic atlantoaxial subluxation. It may occur secondary to head and neck infection or following routine otolaryngological procedures. The underlying mechanism is unknown but haematogenous spread of infection from the posterior pharynx to the cervical spine with hyperemia and abnormal relaxation of the atlantoaxial ligaments is a widely accepted theory [2].

High-risk patients for developing atlantoaxial subluxation are children with Down's syndrome because of greater laxity of atlantoaxial ligaments and increased atlantoaxial space. Other high-risk groups include Klippel–Feil syndrome, osteogenesis imperfecta, neurofibromatosis and any syndromes associated with spinal instability. Nevertheless, atlantoaxial subluxation may also occur in non-syndromic cases with no underlying spinal laxity and early diagnosis and neurosurgical consultation is required to prevent significant morbidity and/or mortality.

2. Methods

A Medline search was performed using the terms Grisel's syndrome, spontaneous atlantoaxial subluxation, la maladie de Grisel, head, neck, ear, nose and throat. A systematic review of the English and non-English literature was performed. Single case reports and case series of both adult and pediatric cases were included. Papers not focusing on Grisel's syndrome but on other postoperative complications of head and neck procedures were excluded. Trauma cases were not included as, by definition, Grisel's syndrome refers to non-traumatic atlantoaxial subluxation.

Papers describing variants but not true atlantoaxial subluxation were excluded. Cases where etiology and/or treatment plan were unclear were also excluded.

3. Results

Seventy-one papers have been published from 1950 to 2006, of which 48 fulfilled our inclusion criteria (Table 1). There were 29 single case reports and 19 case series. The rest of the papers were excluded because the focus was on complications of pediatric adenotonsillectomy in general with no emphasis on Grisel's syndrome. One paper was describing a variant but not true atlantoaxial subluxation.

Overall 103 patients developed AAS of which 7 were post-trauma cases and, therefore excluded. The causes of NAAS are outlined in Table 2. By far the commonest group was infection (48%) followed by postoperative cases (40%). More specifically, out of these postoperative cases (that is the 40%), post-adenotonsillectomy was to blame in 78% of the operations performed, followed by post-pharyngoplasty (15%), post-otoplasty (2.5%), post-tympanomastoidectomy (2.5%) and post-grommets (2.5%). Miscellaneous causes included idiopathic, instillation of eye drops and central line placement. In the infection group, upper respiratory tract infection was to blame for the vast majority of infective cases (83%), followed by retropharyngeal abscess (11%), otitis media (4%) and mumps (2%). The vast majority of the patients reviewed (93%) improved on conservative management whereas the rest required surgery (arthrodesis). Only one case developed a permanent neurodeficit, despite early neurosurgical intervention.

4. Discussion

Despite former belief that the commonest cause for NAAS is postoperative head and neck procedures [2], our results suggest that the commonest cause would be infection followed by otolaryngological operations. Whatever the cause though, early

Table 1 Case studies (in chronological order) investigating the causes of non-traumatic atlantoaxial rotatory subluxation (Grisel's syndrome)

Reference	No. of patients	Etiology
Sipila et al. [5]	2	T
Robinson and De Boer [6]	1	Pharyngoplasty
Hopla et al. [7]	2	First: otitis media, second: choanal atresia
Boiten et al. [8]	2	URTI
Sangermani et al. [9]	1	TA
Robertson et al. [10]	1	RPA
Antoniuk [11]	1	T
Mathern and Batzdorf [12]	2	URTI
Eadie et al. [13]	2	Pharyngoplasty
Singer [14]	2	TA
Litman and Perkins [15]	1	Tympanomastoidectomy& trisomy21
Prado et al. [16]	1	RPA
Samuel et al. [17]	3	TA
Baker et al. [18]	2	TA
Nucci et al. [19]	1	Instilling eye drops& Downs syndrome
Welinder et al. [20]	1	RPA
Onerci et al. [21]	1	TA
Kelly et al. [22]	1	Bilateral otoplasty
*Subach et al. [23]	20	7 URTI, 4 TA, 4 trauma, 5 idiopathic
Bedi et al. [24]	1	TA
Dasen [25]	1	T
Brisson [26]	1	Central line placement
Cunnington and Mongia [27]	1	Pharyngeal abscess
Al-Jishi and Sreekantaswamy [28]	1	Primary dystonia
Meek et al. [29]	1	Pharyngoplasty and grommets in cleft palate case
Kraft and Tschopp [30]	3	A
Holcomb et al. [31]	4	URTI, TA
*Martinez-Lage et al. [32]	4	3 trauma, 1 OM
Guleryuz et al. [33]	1	URTI
Gourin et al. [34]	1	RPA
Okada et al. [35]	1	Mumps
Mezue et al. [36]	13	URTI
Kasten et al. [37]	1	URTI
Tschopp [38]	3	Monopolar electrocautery and A
Ugur et al. [39]	2	URTI
Lehtinen et al. [40]	1	URTI
Yu et al. [41]	1	TA
Fernandez-Cornejo et al. [42]	4	URTI
Hirth and Welkoborsky [43]	2	T
Feldmann et al. [44]	2	TA
Isern et al. [45]	2	Pharyngoplasty
Wurm et al. [46]	1	URTI
Battiata and Pazos [47]	1	TA
Panopalis et al. [48]	1	URTI
Bocciolini et al. [49]	1	A
Galer et al. [50]	1	URTI

The papers marked with an asterisk (*) feature mixed cases of traumatic and non-traumatic atlantoaxial rotatory subluxation. T: Tonsillectomy; A: adenoidectomy; TA: adenotonsillectomy; URTI: upper respiratory tract infection; RPA: retropharyngeal abscess; OM: otitis media.

identification of the atlantoaxial subluxation and – most importantly – neurosurgical consultation is important to prevent significant neurological deficit. Rinaldo et al. concluded that a significant 15% of patients may develop permanent neurological sequelae following NAAS [3].

Diagnosis of NAAS should be based on a high index of suspicion especially for the “non-high risk” groups. Post-tonsillectomy neck pain and torticollis are early signs and should not be attributed to the usual “post-tonsillectomy malaise and pain”. A tender C2 spinous process strongly

Table 2 Causes of Grisel's syndrome

Infection	48%
URTI	83%
RPA	11%
OM	4%
Mumps	2%
Postoperative cases	31%
Adenotonsillectomy	78%
Pharyngoplasty	15%
Tympanomastoidectomy	2.5%
Otoplasty	2.5%
Grommets	2.5%
Miscellaneous (very rare)	12%
Idiopathic	
Central line placement	
Instilling eye drops	

indicates atlantoaxial subluxation. Computerized tomography and magnetic resonance imaging are both excellent diagnostic tools to assess deep neck space infections and the condition/state of the bony and the ligamentous components of the cervical spine. Inflammatory markers are non-specific.

The vast majority of the cases included in the current review did well with conservative management including bedrest, muscle relaxants, cervical traction and soft or hard collar. Early broad spectrum antibiotics are necessary to prevent spread of infection.

Fielding et al. proposed a grading scale for atlantoaxial deformities ranging from type I, a simple fixed deformity to type IV, a serious posterior displacement leading – if untreated – to significant neurosequelae, including quadriplegia and even death [4]. The higher the grade, the more possible the need for surgery. Neurosurgical intervention usually involves arthrodesis/spondylodesis.

5. Conclusion

Non-traumatic atlantoaxial subluxation is a rare but dangerous complication that can often go unnoticed in its early phase and can be a major cause of morbidity and mortality following infection, head and neck procedures and/or any head and neck interventions. Early recognition of any cervical complications following routine otolaryngological operations together with early neurosurgical consultation is mandatory to prevent devastating consequences. The majority of NAAS patients do well if diagnosed early.

References

- [1] P. Grisel, Enucleation de l'atlas et torticollis nasopharyngien, *Presse Med* 59 (78) (1951) 1647–1648.
- [2] G.T. Richter, C.M. Bower, Cervical complications following routine tonsillectomy and adenoidectomy, *Curr. Opin. Otolaryngol. Head Neck Surg.* 14 (6) (2006) 375–380.
- [3] A. Rinaldo, V. Mondin, C. Suarez, E.M. Genden, A. Ferlito, Grisel's syndrome in head and neck practice, *Oral Oncol.* 41 (10) (2005) 966–970.
- [4] J.W. Fielding, R.J. Hawkins, R.N. Hensinger, W.R. Francis, Atlantoaxial rotary deformities, *Orthop. Clin. North Am.* 9 (4) (1978) 955–967.
- [5] P. Sipila, A. Palva, M. Sorri, K. Ojala, Atlantoaxial subluxation. An unusual complication after local anesthesia for tonsillectomy, *Arch. Otolaryngol.* 107 (Mar (3)) (1981) 181–182.
- [6] P.H. Robinson, A. De Boer, La maladie de Grisel: a rare occurrence of "spontaneous" atlanto-axial subluxation after pharyngoplasty, *Br. J. Plast. Surg.* 34 (3) (1981) 319–321.
- [7] D.M. Hopla, J.M. Mazur, R.M. Bass, Cervical vertebrae subluxation, *Laryngoscope* 93 (9) (1983) 1155–1159.
- [8] J. Boiten, G. Hageman, R. de Graaff, The conservative treatment of patients presenting with Grisel's syndrome, *Clin. Neurol. Neurosurg.* 88 (2) (1986) 95–99.
- [9] R. Sangermani, F. Micheloni, A. Partesana, C. Cammarata, P. Pacilli, F. Lucchini, Grisel's syndrome description of a case of spontaneous subluxation of the atlas, *Pediatr. Med. Chir.* 8 (5) (1986) 735–736.
- [10] S. Robertson, M.L. Pinstein, D.G. LaVelle, Non-traumatic atlantoaxial subluxation in an adult secondary to retropharyngeal abscess, *Orthopedics* 10 (11) (1987) 1545–1547.
- [11] M.I. Antoniuk, Grisel's disease after tonsillectomy, *Vestn Otorinolaringol.* (1) (1988) 72–73.
- [12] G.W. Mathern, U. Batzdorf, Grisel's syndrome. Cervical spine clinical, pathologic, and neurologic manifestations, *Clin. Orthop. Relat. Res.* (244) (1989) 131–146.
- [13] P.A. Eadie, R. Moran, E.E. Fogarty, G.E. Edwards, Rotatory atlantoaxial subluxation following pharyngoplasty, *Br. J. Plast. Surg.* 42 (6) (1989) 722–723.
- [14] J.I. Singer, Evaluation of the patient with neck complaints following tonsillectomy or adenoidectomy, *Pediatr. Emerg. Care.* 8 (5) (1992) 276–279.
- [15] R.S. Litman, F.M. Perkins, Atlantoaxial subluxation after typanomastoidectomy in a child with trisomy 21, *Otolaryngol. Head Neck Surg.* 110 (6) (1994) 584–586.
- [16] V.M. Prado, L.D. Giardini, M. Garcia, R.M. DeSousa, G.G. Formigoni, Retropharyngeal abscess after adenoidectomy, *Ear Nose Throat J.* 74 (1) (1995) 54–55.
- [17] D. Samuel, D.M. Thomas, P.A. Tierney, K.S. Patel, Atlantoaxial subluxation (Grisel's syndrome) following otolaryngological diseases and procedures, *J. Laryngol. Otol.* 109 (10) (1995) 1005–1009.
- [18] L.L. Baker, C.M. Bower, C.M. Glasier, Atlanto-axial subluxation and cervical osteomyelitis: two unusual complications of adenoidectomy, *Ann. Otol. Rhinol. Laryngol.* 105 (4) (1996) 295–299.
- [19] P. Nucci, M. de Pellegrin, R. Brancato, Atlantoaxial dislocation related to instilling eyedrops in a patient with Down's syndrome, *Am. J. Ophthalmol.* 122 (6) (1996) 908–910.
- [20] N.R. Welinder, P. Hoffmann, S. Hakansson, Pathogenesis of non-traumatic atlanto-axial subluxation (Grisel's syndrome), *Eur. Arch. Otorhinolaryngol.* 254 (5) (1997) 251–254.

- [21] M. Onerci, O. Ogretmenoglu, O.E. Ozcan, Atlantoaxial subluxation after tonsillectomy and adenoidectomy, *Otolaryngol. Head Neck Surg.* 116 (2) (1997) 271–273.
- [22] E.J. Kelly, K.J. Herbert, E.J. Crotty, T.P. O'Connor, Atlantoaxial subluxation after otoplasty, *Plast. Reconstr. Surg.* 102 (2) (1998) 543–544.
- [23] B.R. Subach, M.R. McLaughlin, A.L. Albright, I.F. Pollack, Current management of pediatric atlantoaxial rotatory subluxation, *Spine* 23 (20) (1998) 2174–2179.
- [24] H.S. Bedi, R.D. Angliss, S.A. Williams, D.P. Connelly, Torticollis following adenotonsillectomy, *Aust. N Z J. Surg.* 69 (2) (1999) 163–164.
- [25] K.R. Dasen, Atlantoaxial rotatory subluxation after a pediatric tonsillectomy, *Anesth. Analg.* 89 (4) (1999) 917–919.
- [26] P. Brisson, H. Patel, R. Scorpio, N. Feins, Rotary atlanto-axial subluxation with torticollis following central-venous catheter insertion, *Pediatr. Surg. Int.* 16 (5–6) (2000) 421–423.
- [27] P. Cunnington, S. Mongia, Pharyngeal abscess in a small infant presenting as upper airway obstruction and atlantoaxial subluxation, *Anaesthesia* 55 (9) (2000) 927–928.
- [28] A. Al-Jishi, Sreekantaswamy, Dystonia associated with atlantoaxial subluxation, *Clin. Neurol. Neurosurg.* 102 (4) (2000) 233–235.
- [29] M.F. Meek, R.A. Hermens, P.H. Robinson, La maladie de Grisel: spontaneous atlantoaxial subluxation, *Cleft Palate Craniofac J.* 38 (3) (2001) 268–270.
- [30] M. Kraft, K. Tschopp, Evaluation of persistent torticollis following adenoidectomy, *J. Laryngol. Otol.* 115 (8) (2001) 669–672.
- [31] J.D. Holcomb, D.M. Jaffe, J.H. Greinwald Jr., N.M. Bauman, R.J. Smith, Nontraumatic atlantoaxial rotary subluxation in the pediatric otolaryngology patient. A report of four cases, *Ann. Otol. Rhinol. Laryngol.* 110 (12) (2001) 1137–1140.
- [32] J.F. Martinez-Lage, M. Martinez Perez, V. Fernandez Cornejo, M. Poza, Atlanto-axial rotatory subluxation in children: early management, *Acta Neurochir. (Wien)* 143 (12) (2001) 1223–1228.
- [33] A. Guleryuz, C. Bagdatoglu, M.N. Duce, D.U. Talas, H. Celikbas, T. Koksel, Grisel's syndrome, *J. Clin. Neurosci.* 9 (1) (2002) 81–84.
- [34] C.G. Gourin, B. Kaper, W.A. Abdu, J.O. Donegan, Nontraumatic atlanto-axial subluxation after retropharyngeal cellulitis: Grisel's syndrome, *Am. J. Otolaryngol.* 23 (1) (2002) 60–65.
- [35] Y. Okada, N. Fukasawa, T. Tomomasa, Y. Inoue, A. Morikawa, Atlanto-axial subluxation (Grisel's syndrome) associated with mumps, *Pediatr. Int.* 44 (2) (2002) 192–194.
- [36] W.C. Mezue, Z.M. Taha, E.M. Bashir, Fever and acquired torticollis in hospitalized children, *J. Laryngol. Otol.* 116 (4) (2002) 280–284.
- [37] P. Kasten, J. Zeichen, T. Gosling, C. Krettek, Grisel syndrome—a trauma surgery rarity, *Unfallchirurg* 105 (6) (2002) 565–568.
- [38] K. Tschopp, Monopolar electrocautery in adenoidectomy as a possible risk factor for Grisel's syndrome, *Laryngoscope* 112 (8 Pt 1) (2002) 1445–1449.
- [39] H.C. Ugur, S. Caglar, A. Unlu, A. Erdem, Y. Kanpolat, Infection-related atlantoaxial subluxation in two adults: Grisel syndrome or not? *Acta Neurochir. (Wien)* 145 (1) (2003) 69–72.
- [40] P. Lehtinen, O. Ruuskanen, P. Sonninen, An unusual life threatening cause of torticollis in a child, *Arch. Dis. Child.* 88 (4) (2003) 349.
- [41] K.K. Yu, D.R. White, M.C. Weissler, H.C. Pillsbury, Nontraumatic atlantoaxial subluxation (Grisel syndrome): a rare complication of otolaryngological procedures, *Laryngoscope* 113 (6) (2003) 1047–1049.
- [42] V.J. Fernandez Cornejo, J.F. Martinez-Lage, C. Piqueras, A. Gelabert, M. Poza, Inflammatory atlanto-axial subluxation (Grisel's syndrome) in children: clinical diagnosis and management, *Childs Nerv. Syst.* 19 (5–6) (2003) 342–347.
- [43] K. Hirth, H.J. Welkoborsky, Grisel's syndrome following ENT-surgery: report of two cases, *Laryngorhinotologie* 82 (11) (2003) 794–798.
- [44] H. Feldmann, E.F. Meister, K. Kuttner, From the expert's office. Atlanto-axial subluxation with spastic torticollis after adenoidectomy resp. tonsillectomy in Rose position—malpractice of the surgeon or the anaesthesiologist? *Laryngorhinotologie* 82 (11) (2003) 799–804.
- [45] A.E. Isern, A. Ohlin, L.G. Stromblad, I.R. Johnell, M. Becker, H. Svensson, Grisel syndrome after velopharyngoplasty, *Scand. J. Plast. Reconstr. Surg. Hand Surg.* 38 (1) (2004) 53–57.
- [46] G. Wurm, M. Aichholzer, K. Nussbaumer, Acquired torticollis due to Grisel's syndrome: case report and follow-up of non-traumatic atlantoaxial rotatory subluxation, *Neuropediatrics* 35 (2) (2004) 134–138.
- [47] A.P. Battiata, G. Pazos, Grisel's syndrome: the two-hit hypothesis—a case report and literature review, *Ear Nose Throat J.* 83 (8) (2004) 553–555.
- [48] P. Panopalis, S. Christopoulos, M. Churchill-Smith, J. Chankowsky, H.A. Menard, Grisel' syndrome: non-traumatic subluxation of the atlantoaxial joint, *J. Rheumatol.* 32 (8) (2005) 1619.
- [49] C. Bocciolini, D. Dall'Olivo, E. Cunsolo, P.P. Cavazzuti, P. Laudadio, Grisel's syndrome: a rare complication following adenoidectomy, *Acta Otorhinolaryngol. Ital.* 25 (4) (2005) 245–249.
- [50] C. Galer, E. Holbrook, J. Treves, D. Leopold, Grisel's syndrome: a case report and review of the literature, *Int. J. Pediatr. Otorhinolaryngol.* 69 (12) (2005) 1689–1692.