CASE IN POINT

Mild Grisel Syndrome: Expanding the Differential for Posttonsillectomy Adenoidectomy Symptoms

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Background: Tonsillectomy with or without adenoidectomy is the second most common pediatric surgical procedure in the United States with up to 97% performed as an outpatient.[1,2] While it is largely a safe procedure, several complications have been described and are encountered in the emergency department and primary care setting.

Presentation: A 29-month child presented to the emergency department with neck stiffness 10 days after tonsillectomy and adenoidectomy. A computed tomography scan of the neck limited by motion artifact was unrevealing, but a consult to the pediatric otolaryngologist generated concern for Grisel syndrome, the atraumatic rotary subluxation of the atlantoaxial joint. While surgical intervention can be required, the patient had an uncomplicated clinical course and the anomalous neck posture resolved with time and anti-inflammatories alone.

Conclusions: Keeping a broad differential for posttonsillectomy and adenoidectomy patient concerns is important for the clinician. Serious, life-threatening complications can arise from Grisel syndrome while good functional outcomes can be achieved with timely and appropriate treatment.

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Tonsillectomy with or without adenoidectomy (T&A) is the second most common pediatric surgical procedure in the United States.[1] It is most often performed during childhood between 5 and 8 years of age with a second peak observed between 17 and 21 years of age in the adolescent and young adult populations.[2] While recurrent tonsillitis has been traditionally associated with tonsillectomy, sleep disordered breathing with obstructive sleep apnea is now the primary indication for the procedure.[1]

Up to 97% of T&As are performed as an outpatient same-day surgery not requiring inpatient admission.[2] Although largely a safe and routinely performed surgery, several complications have been described. Due to the outpatient nature of the procedure, the complications are often encountered in the emergency department (ED) and sometimes in primary care settings. Common complications (outside of the perioperative time frame) include nausea, vomiting, otalgia, odynophagia, infection of the throat (broadly), and hemorrhage; uncommon complications include subcutaneous emphysema, taste disorders, and Eagle syndrome. Some complications are rarer still and carry significant morbidity and even mortality, including mediastinitis, cervical osteomyelitis, and Grisel syndrome.[3] The following case encourages the clinician to expand the differential for a patient presenting after T&A.

CASE PRESENTATION

A child aged < 3 years was brought to the ED by their mother. She reported neck pain and stiffness 10 days after T&A with concurrent tympanostomy tube placement at an outside pediatric hospital. At triage, their heart rate was 94 bpm, temperature was 98.2 °F, respiratory rate, 22 breaths per minute, and oxygen saturation, 97% on room air. The mother of the patient (MOP) had been giving the prescribed oral liquid formulations of ibuprofen and acetaminophen with hydrocodone as directed. No drug allergies were reported, and immunizations were up to date for age. Other medical and surgical history included eczema and remote cutaneous hemangioma resection. The patient lived at home with 2 parents and was not exposed to smoke; their family history was noncontributory.

Since the surgery, the MOP had noticed constant and increasing neck stiffness, specifically with looking up and down but not side to side. She also had noticed swelling behind both ears. She reported no substantial decrease in intake by mouth or decrease in urine or bowel frequency. On review of

systems, she reported no fever, vomiting, difficulty breathing, bleeding from the mouth or nose, eye or ear drainage, or rash.

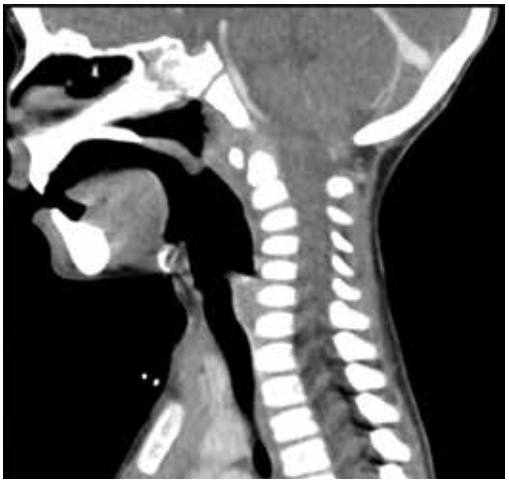
On physical examination, the patient was alert and in no acute distress; active and playful on an electronic device but was notably not moving their head, which was held in a forward-looking position without any signs of trauma. When asked, the child would not flex or extend their neck but would rotate a few degrees from neutral to both sides. Even with moving the electronic device up and down in space, no active neck extension or flexion could be elicited. The examination of the head, eyes, ears, nose, and throat was otherwise only remarkable for palpable and mildly tender postauricular lymph nodes and diffuse erythema in the posterior pharynx. Cardiopulmonary, abdominal, skin, and extremity examinations were unremarkable.

With concern for an infectious process, the physician ordered blood chemistry and hematology tests along with neck radiography. While awaiting the results, the patient was given a weight-based bolus of normal saline, and the home pain regimen was administered. An attempt was made to passively flex and extend the neck as the child slept in their mother's arms, but the patient immediately awoke and began to cry.

All values of the comprehensive metabolic panel were within normal limits except for a slight elevation in the blood urea nitrogen to 21 mg/dL and glucose to 159 mg/dL. The complete blood count was unrevealing. The computed tomography (CT) scan with contrast of the soft tissues of the neck was limited by motion artifact but showed a head held in axial rotation with soft tissue irregularity in the anterior aspect of the adenoids (Figure 1). There was what appeared to be normal lymphadenopathy in the hypopharynx, but the soft tissues were otherwise unremarkable.

The on-call pediatric otolaryngologist at the hospital where the procedure was performed was paged. On hearing the details of the case, the specialist was concerned for Grisel syndrome and requested to see the patient in their facility. No additional recommendations for care were provided; the mother was updated and agreed to transfer.

FIGURE 1 Sagittal Computed Tomography in Soft Tissue Window Showing Artifact in the Hypopharynx



The patient was comfortable and stable with repeat vitals as follows: heart rate, 86 beats per minute, blood pressure, 99/62, temperature, 98.3 °F, respiratory rate, 20 breaths per minute, and oxygen saturation, 99% on room air.

On arrival at the receiving facility, the emergency team performed a history and physical that revealed no significant changes from the initial evaluation. They then facilitated evaluation by the pediatric otolaryngologist who conducted a more directed physical examination. Decreased active and passive range of motion (ROM) of the neck without rotatory restriction was again noted. They also observed scant fibrinous exudate within the oropharynx and tonsillar fossa, which was normal in the setting of the recent surgery. They recommended additional analgesia with intramuscular ketorolac, weight-based dosing at 1 mg/kg.

With repeat examination after this additional analgesic, ROM of the neck first passive then active had improved. The patient was then discharged to follow up in the coming days with instructions to continue the pain and anti-inflammatory regimen. They were not started on an antibiotic at that time nor were they placed in a cervical collar. At the follow-up, the MOP reported persistence of neck stiffness for a few days initially but then observed slow improvement. By postoperative day 18, the stiffness had resolved. No other follow-up or referrals

TABLE Fielding and Hawkins Classification for Grisel Syndrome

Class	Dislocation and Displacement of Atlas	Atlantodens Interval, mm	Neurologic Impairment
I	Rotary without displacement	Minimal	No
II	Rotary with anterior	≤ 5	No
III	Rotary with anterior	> 5	Yes or no
IV	Rotary with posterior	Any	Yes

related to this issue were readily apparent in review of the patient's health record.

DISCUSSION

Grisel syndrome is the atraumatic rotary subluxation of the atlantoaxial joint, specifically, the atlas (C1 vertebra) rotates to a fixed, nonanatomic position while the axis (C2 vertebra) remains in normal alignment in relation to the remainder of the spinal column. The subluxation occurs in the absence of ligamentous injury but is associated with an increase in ligamentous laxity.[4] The atlas is a ring-shaped vertebra with 2 lateral masses connected by anterior and posterior arches; it lacks a spinous process unlike other vertebrae. It articulates with the skull by means of the 2 articular facets on the superior aspect of the lateral masses. Articulation with the axis occurs at 3 sites: 2 articular facets on the inferior portion of the lateral masses of the atlas and a facet for the dens on the posterior portion of the anterior arch. The dens projects superiorly from the body of the axis and is bound posteriorly by the transverse ligament of the atlas.[5]

The degree of subluxation seen in Grisel syndrome correlates to the disease severity and is classified by the Fielding and Hawkins (FH) system (Table). This system accounts for the distance from the atlas to the dens (atlantodens interval) and the relative asymmetry of the atlantoaxial joint.[6] In a normal adult, the upper limit of normal for the atlantodens interval is 3 mm, whereas this distance increases to 4.5 mm for the pediatric population.[7] Type I (FH-I) involves rotary subluxation alone without any increase in the atlantodens interval; in FH-II, that interval has increased from normal but to no more than 5 mm. FH-I and FH-II are the most encountered and are not associated with neurologic impairment. In FH-III, neurologic deficits can be present, and the atlantodens interval is increased to > 5 mm. Different from FH-II and FH-III in which anterior dislocation of the atlas with reference to the dens is observed, FH-IV involves a rotary movement of the atlas with concurrent posterior displacement and often involves spinal cord compression.[6]

Subluxation and displacement without trauma are key components of Grisel syndrome. The 2-hit hypothesis is often used to explain how this can occur, ie, 2 anomalies must be present simultaneously for this condition to develop. First, the laxity of the transverse ligament, the posterior wall of the dens, and other atlantoaxial ligaments must be increased. Second, an asymmetric contraction of the deep erector muscles of the neck either abruptly or more insidiously rotate and dislocate the atlas.[8] The pathophysiology is not exactly understood, but the most commonly held hypothesis describes contiguous spread of infection or inflammatory mediators from the pharynx to the ligaments and muscles described.[6]

Spread could occur via the venous system. The posterior superior pharyngeal region is drained by the periodontoidal venous plexus; the connections here with the pharyngovertebral veins allow for the embolization of infectious or other proinflammatory material to the prevertebral fascia. These emboli induce fasciitis and subsequent aberrant relaxation of the ligaments. In reaction to the inflammation or increased laxity, contiguous muscles of the deep neck contract and freeze the joint out of anatomic alignment.[4]

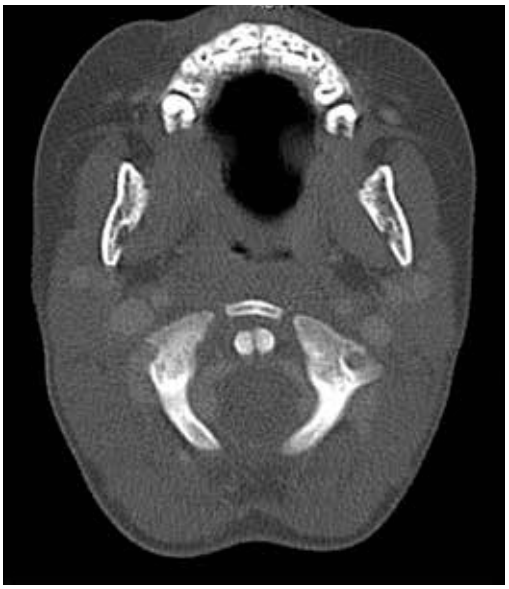
The abnormal alignment is apparent grossly as torticollis. Most broadly, torticollis describes an anomalous head posture due to involuntary muscle contractions of neck muscles and specifically describes

chin deviation to the side. The antecollis and retrocollis subtypes of torticollis describe forward flexion and backward extension of the neck, respectively.[7] Torticollis (broadly) is the most frequently reported condition of those found to have Grisel syndrome (90.7%); other common presenting conditions include neck pain (81.5%) and neck stiffness (31.5%). Fever is found in only 27.8% of cases. Pediatric patients (aged ≤ 12 years) are the most commonly affected, accounting for 87% of cases with an observed 4:1 male to female predominance.[7,8] Symptoms begin most often within the first week from the inciting event in 85% of the cases.[8] Head and neck surgery precedes up to 67% of cases, and infectious etiologies largely account for the remaining cases.[7] Of the postsurgical cases, 55.6% had undergone T&A.[8]

Although anomalous head posture or neck stiffness following T&A would be of great clinic concern for Grisel syndrome, radiographic studies play a confirmatory role. CT scan is used to evaluate the bony structures, with 3D reconstruction of the cervical spine being most useful to determine the presence and degree of subluxation.[8] Magnetic resonance imaging also aids in diagnosis to evaluate ligamentous structures in the area of concern as well as in the evaluation of spinal cord compression.[6] Laboratory tests are largely unhelpful in making or excluding the diagnosis.[8]

If Grisel syndrome is suspected, both the original surgeon (if preceded by surgery) and the neurosurgical team should be consulted. Although no widely adopted guidelines exist for the management of this rare disease, general practice patterns have emerged with the degree of intervention predictably correlating to disease severity. FH-I is usually treated with nonsteroidal anti-inflammatory drugs and muscle relaxants with or without a soft cervical collar. For FH-II, closed reduction and immobilization in a stiff cervical collar is recommended. If no neurologic defect is present, FH-III is treated with bed rest, a period of inline cervical traction, and subsequent immobilization. FH-III with neurologic sequelae and all FH-IV necessitate emergent neurosurgical consultation.[4] Surgical intervention is a last resort but is required in up to 24.1% of cases.[8]

FIGURE 2 Axial Computed Tomography in Bone Window



Antibiotic therapy is not routinely given unless clear infectious etiology is identified. No standard antibiotic regimen exists, but coverage for typical upper respiratory pathogens likely suffices. Empiric antibiotic therapy is not recommended for all causes of Grisel syndrome, ie, when the underlying cause is not yet elucidated.[6] One case of Grisel syndrome occurring in the setting of cervical osteomyelitis has been described, though, and required prolonged IV antibiotics.[3] Physical therapy is recommended as adjunct with no limitations for range of motion save for that of the patient's individual pain threshold.[4]

Possibly attributable to waxing and waning ligamentous laxity and strength of the neck muscle contraction, the atlantodens interval and the degree of subluxation can change, making Grisel syndrome dynamic. As such, the FH classification can change, necessitating more or less aggressive therapy. A neurologic evaluation is recommended at least every 2 weeks after the diagnosis is made. If initial identification or recognition of known disease progression is delayed, serious complications can develop. Acutely, spinal cord compression can lead to quadriplegia and death; more insidious complications include reduced neck mobility, dysphonia, and dysphagia.[4] As serious, life-threatening

complications can arise from Grisel syndrome while good functional outcomes can be achieved with timely and appropriate treatment, the clinician should be inspired to have a high clinical suspicion for this syndrome given the right context.

CONCLUSIONS

The patient experienced a desirable outcome with minimal, conservative treatment. As such, the pathology in this case was likely attributed to the mildest form of Grisel syndrome (FH-I). The follow-up was reassuring as well, revealing no worsening or progression of symptoms. The initial evaluation in this case was limited by the inadequacy of the CT scan. Motion artifact in the pharynx prevented the definite exclusion of deep space infection, while the rotation of the head in combination with motion artifact in the cranial-most portions of the vertebral column made determining alignment difficult. One clear axial image, though, does show rotation of the atlas (Figure 2). The uncertainty at the end of our workup prompted surgical consultation, not, admittedly, concern for Grisel syndrome. Awareness of this disease entity is nevertheless important and clinically relevant. Early identification and treatment is associated with decreased morbidity and improvement in long-term functional outcomes.[6] Despite its rarity, the clinician should consider Grisel syndrome in any pediatric patient presenting with neck stiffness following the commonly performed T&A.

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The opinions expressed herein are those of the authors and do not necessarily reflect those of *Federal Practitioner,* Frontline Medical Communications Inc., the US Government, or any of its agencies.

Ethics and consent

The authors report that written consent was not obtained from the guardian of the patient. Details have been changed to avoid identification.

References

1. Ramos SD, Mukerji S, Pine HS. Tonsillectomy and adenoidectomy. *Pediatr Clin North Am.* 2013;60(4):793-807. doi:10.1016/j.pcl.2013.04.015
2. Stoner MJ, Dulaurier M. Pediatric ENT emergencies. *Emerg Med Clin North Am.* 2013;31(3):795-808. doi:10.1016/j.emc.2013.04.005
3. Leong SC, Karoos PD, Papouliakos SM, et al. Unusual complications of tonsillectomy: a systematic review. *Am J Otolaryngol.* 2007;28(6):419-422. doi:10.1016/j.amjoto.2006.10.016
4. Fath L, Cebula H, Santin MN, Cocab A, Debrya C, Proustb F. The Grisel's syndrome: a non-traumatic subluxation of the atlantoaxial joint. *Neurochirurgie.* 2018;64(4):327-330. doi:10.1016/j.neuchi.2018.02.001
5. Moore K, Agur A, Dalley A. *Essential Clinical Anatomy.* 5th ed. Baltimore: Lippincott, Williams, and Wilkins; 2015:282-287.
6. Spennato P, Nicosia G, Rapanà A, et al. Grisel syndrome following adenoidectomy: surgical management in a case with delayed diagnosis. *World Neurosurg.* 2015;84(5):1494.e7-e12.
7. Anania P, Pavone P, Pacetti M, et al. Grisel syndrome in pediatric age: a single-center Italian experience and review of the literature. *World Neurosurg.* 2019;125:374-382. doi:10.1016/j.wneu.2019.02.035
8. Aldriweesh T, Altheyab F, Alenezi M, et al. Grisel's syndrome post otolaryngology procedures: a systematic review. *Int J Pediatr Otorhinolaryngol.* 2020;137:110-125. doi:10.1016/j.ijporl.2020.110225