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What is This?
Velopharyngeal Incompetence as a Complication of Grisel Syndrome

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Keywords
Grisel syndrome, torticollis, cranial nerve palsy, neurologic complication, atlantoaxial rotatory subluxation, dysphagia, dysphonia

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Atraumatic atlantoaxial subluxation, also known as Grisel syndrome, is a well-characterized but rare entity presenting with symptomatic torticollis in the setting of underlying cervical inflammation, typically due to surgery or infection.¹ Neurologic complications are uncommon and generally result from impingement on the spinal cord.² We present a case of Grisel syndrome in an 8-year-old girl with the previously unreported complication of velopharyngeal incompetence.

Case Report

An 8-year-old girl was evaluated at our institution for torticollis, dysphagia, nasal regurgitation, and voice change. Six weeks prior, she awoke with her head turned to the right and limited neck range of motion. Her pediatrician prescribed physical therapy, baclofen, and diazepam, which did not resolve the symptoms. Two days prior to presentation, she developed a “nasal” quality to her voice, difficulty swallowing solids, and nasal regurgitation of liquids. She was seen in the emergency department and was admitted to the pediatrics service.

Computed tomography demonstrated atlantoaxial rotatory subluxation without significant anterior displacement (Figure 1). The otolaryngology service was consulted for evaluation of dysphagia and voice change.

Physical examination demonstrated lack of elevation of her right soft palate, uvular deviation to the left, hypernasal voice, limited neck range of motion, and head flexed and turned to the right. Flexible fiberoptic laryngoscopy showed failure of closure of the velopharyngeal port on the right but was otherwise normal, with normal vocal cord mobility bilaterally.

Neurosurgery was immediately consulted, and treatment with manual traction, diazepam, and a cervical collar was initiated. Her dysphagia and hypernasal speech quickly resolved. On 4-week follow-up in our clinic, she had symmetric and full palatal elevation bilaterally.

Discussion

Atraumatic atlantoaxial rotatory subluxation (Grisel syndrome) is an uncommon entity that typically presents following surgery or infection of the head and neck, more frequently in children.¹ The condition is caused by lax spinous ligaments due to an inflammatory state.³ Tonsillectomy and adenoidectomy are frequently cited as inciting factors, but a wide variety of head and neck procedures have been associated with this syndrome; otolaryngologists should have a high index of suspicion for this condition following any head and neck procedure.³ This case is unusual, although not unprecedented, in the lack of an inciting factor in the development of her subluxation. In a review by Karkos et al⁴ of 96 patients with atraumatic atlantoaxial subluxation described in prior publications, 12% had neither antecedent infection nor surgery.

Fielding and Hawkins⁴ described 4 types of atlantoaxial rotatory subluxation. Type I, as in this case, is the most common and consists of rotatory subluxation with less than 3 mm of anteroposterior displacement. Type II describes subluxation between 3 and 5 mm. Type III is greater than 5 mm displacement and is considerably more likely to lead to symptoms, and type IV has rotatory subluxation and posterior displacement and has substantial potential to lead to neurologic sequelae.

Neurologic sequelae have been described to occur in 15% of patients with atlantoaxial rotatory subluxation, with death due to respiratory failure occurring in 6% of an early series.² Literature review identified no previous publications describing a patient presenting with isolated cranial nerve weakness. In this case, our patient’s palatal weakness, manifesting in hypernasal speech and difficulty swallowing, likely represented an isolated deficit of a branch of cranial nerve X. Fortunately, this deficit resolved with successful

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treatment of her subluxation, supporting the subluxation as the cause of the cranial nerve weakness.

Prompt diagnosis is crucial and can often be made by history and physical alone, particularly if the patient has had recent head and neck surgery or infection and has the classic physical signs of a palpable C2 spinous process, torticollis, and ipsilateral sternocleidomastoid spasm. Computed tomography should be obtained to confirm the diagnosis and define the extent of subluxation. Once the diagnosis has been made, neurosurgical consultation is essential, as prompt management may prevent complications.

Treatment, as in this case, is typically conservative. Initial management is traction coupled with muscle relaxants and analgesics. Patients who fail this may require halo fixation or cervical spine fixation. In this case, failure to initiate traction upon her initial presentation may have allowed progression of neurologic sequelae.

As per the policy of the Research Institute at Nationwide Children’s Hospital, this case report is exempt from institutional review.

Author Contributions

Benjamin C. Tweel, data acquisition and interpretation, drafting of article, final approval; Charles Elmaraghy, data acquisition and interpretation, drafting of article, final approval.

Disclosures

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