Grisel Syndrome Following Adenoidectomy: Surgical Management in a Case with Delayed Diagnosis

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Key words
- Atlantoaxial fixation
- Atlantoaxial instability
- Atlantoaxial rotatory subluxation
- Ear, nose, and throat complication

Abbreviations and Acronyms
CT: Computed tomography
ENT: Ear, nose, and throat
FFE: Fast field echo
MEP: Motor evoked potentials
MIP: Maximum intensitive projection
MPR: Multiplanar reconstruction
MRI: Magnetic resonance imaging
SSEP: Somatosensorial evoked potentials
WI: Weighted images

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BACKGROUND: **Grisel syndrome is a nontraumatic rotatory subluxation of the atlantoaxial joint, following nasopharyngeal inflammation or ear, nose, and throat (ENT) procedures. The syndrome should be suspected in cases of persistent neck pain and stiffness, especially after ENT surgical procedures. The primary treatment of early detected Grisel syndrome is conservative. If conservative treatment fails to achieve a stable reduction or it is followed by neurologic symptoms, arthrodesis of the first and second cervical vertebrae is indicated. We report the case of a 9-year-old boy who developed Grisel syndrome after adenoidectomy and was treated with C1-C3 internal fixation and fusion.**

CASE DESCRIPTION: **A 9-year-old boy was referred to our hospital with a 3-month history of painful torticollis, which appeared 4 days after adenoidectomy. The patient underwent a neuroimaging study that documented the presence of atlantoaxial rotatory subluxation. The patient underwent C1-C3 internal fixation and fusion, using lateral masses and laminar and pars interarticularis screws. On the third postoperative day he was mobilized with a rigid collar. Postoperative computed tomography scans showed the resolution of rotational deformity and a solid fusion.**

CONCLUSION: **Early treatment of Grisel syndrome is of utmost importance to avoid neurologic complications and surgical intervention. In a patient with torticollis following ENT procedures, Grisel syndrome should be always suspected. In case of failure of conservative treatment or in case of delayed diagnosis, rigid C1-C2 or C1-C2-C3 fixation is a straightforward and valid surgical technique, even in children, because it provides immediate spinal stability in all planes at the atlantoaxial complex, avoiding the need for prolonged rigid external bracing.**

INTRODUCTION

Grisel syndrome is a spontaneous rotatory subluxation of the atlantoaxial joint not associated with trauma or bone disease, found primarily in children (7). It may occur after nasopharyngeal inflammation or ear, nose, and throat (ENT) procedures such as tonsillectomy, adenoidectomy, and mastoidectomy. It should be suspected in cases of persistent neck pain and stiffness, especially after ENT procedures. Radiographs and computed tomography (CT) scans of the cervical spine confirm the diagnosis. Early management, consisting of closed reduction and cervical immobilization and medical treatment, is the key factor for a satisfactory outcome (8). We report the case of a 9-year-old boy who developed Grisel syndrome after adenoidectomy. The radiological diagnosis and the surgical treatment are hereby discussed.

CASE REPORT

An Italian 9-year-old boy underwent adenoidectomy and bilateral tonsillar reduction, under general anaesthesia. The procedure was uneventful, and the patient was discharged home the day after surgery. On the fourth postoperative day, painful torticollis occurred with mild rotation of the patient's head to the right. For this reason the patient was referred to the emergency department. The physical examination revealed a spasm in the right sternocleidomastoid muscle, and the patient was discharged with a diagnosis of torticollis, recommending medical therapy and neck immobilization with cervical collar, without radiological investigations. The patient was referred to the emergency department three more times because of persistent painful torticollis. Finally, about 2 months after ENT surgery, the patient underwent neuroimaging examinations that documented the presence of an atlantoaxial rotatory subluxation. Therefore the patient was referred to the neurosurgical department. At admission the patient was neurologically intact and exhibited head rotation to the right side and tilt to the left, in slight flexion. Neck

mobility was restricted on each side. Pediatric and ENT consultations showed no signs of inflammation/infection of the upper airways. Hematological and biochemical tests were within the normal range. The patient was initially treated with a cervical collar, antiinflammatory and muscle relaxant drugs for 2 weeks, with no improvement of the clinical and radiological scenario.

Radiological Findings

Radiological evaluation included radiograph of the cervical spine with anteroposterior, lateral, odontoid projections and dynamic flexion-extension study; CT scan with 3-D reconstructions; and magnetic resonance imaging (MRI) of cervical spine. Radiograph revealed the typical head tilt of the atlantoaxial subluxation; reduced mobility during flexion; and no mobility during extension. On the CT scan, the 3-D reconstructions confirmed the atlantoaxial rotatory subluxation with anterolateral dislocation of C1 and showed a clearly widened distance between the atlas and dens, measuring 6.7 mm. The atlas was right rotated 45 degrees with respect to the axis (Figure 1). The T1-weighted MRI showed a widened atlantodental space with heterogeneous hyperintensity in T2-weighted images at this level and interruption of the signal of the transverse ligament on the left (Figure 2). Angio CT was useful to identify the relationship of the vertebral artery with the posterior elements of the upper cervical vertebrae and to exclude anomalies in its course (Figure 3).

Operation

The patient underwent open C1-C3 internal fixation and fusion. Under general anaesthesia, with the patient in prone position and the head fixed in a three-pin headframe, a reduction of the rotatory subluxation was accomplished under fluoroscopy: a gentle stretching of the patient's neck along with a gentle left rotary maneuver was given. During the procedure, an assistant fixed the shoulders of the patient to prevent any motion. Therefore internal rigid fixation was achieved with C1 lateral masses screws (Figure 4A) and C3 pars interarticularis screws (Figure 4C); one intralaminar screw was also placed in the left lamina of C2 starting from the right in order to push the spinous process of C2 to the left to counteract the rotation of the axis (Figure 4B). The system was assembled with lordotic-shaped titanium bars (Figure 4D-G); the fusion was favored with the aid of granules of tricalcium phosphate, positioned following decortication of the laminae and articular processes of the exposed vertebrae (C1-C3).

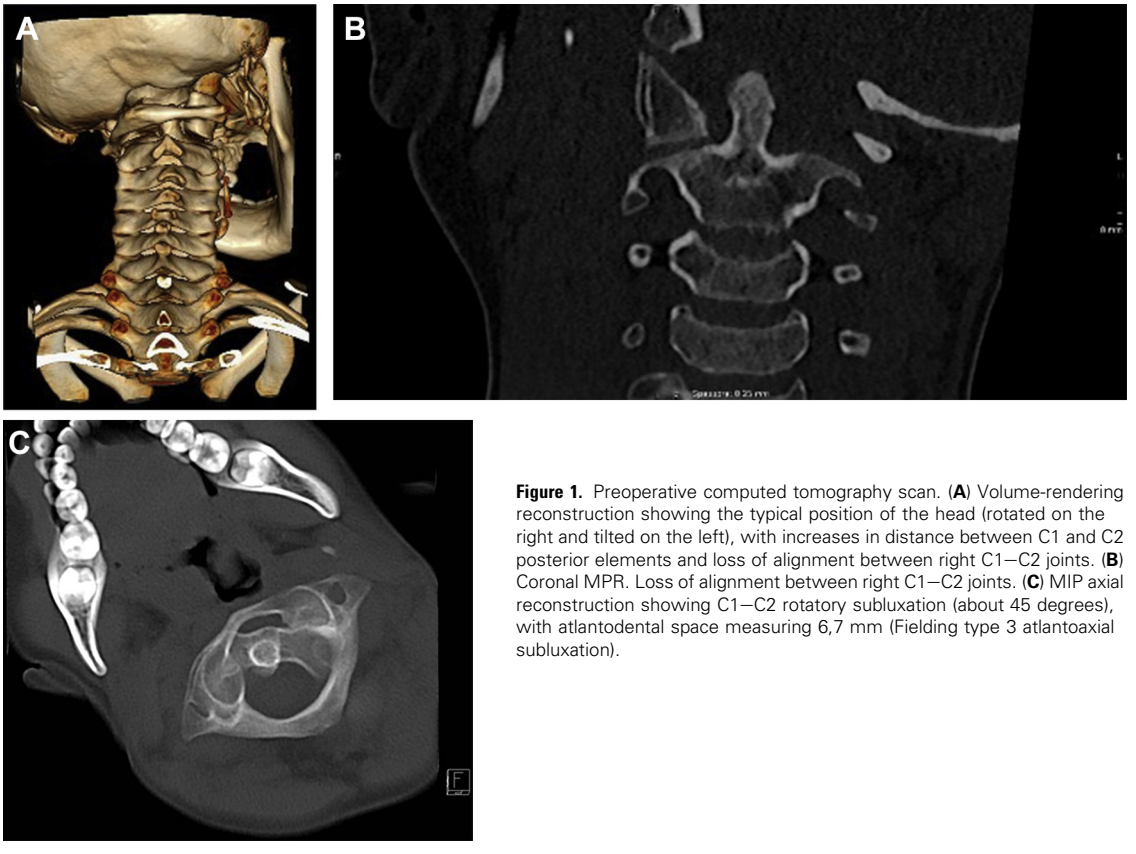

Figure 1. Preoperative computed tomography scan. (**A**) Volume-rendering reconstruction showing the typical position of the head (rotated on the right and tilted on the left), with increases in distance between C1 and C2 posterior elements and loss of alignment between right C1—C2 joints. (**B**) Coronal MPR. Loss of alignment between right C1—C2 joints. (**C**) MIP axial reconstruction showing C1—C2 rotatory subluxation (about 45 degrees), with atlantodental space measuring 6,7 mm (Fielding type 3 atlantoaxial subluxation).