Grisel Syndrome - A case report

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Abstract

Grisel's syndrome (GS) is the non-traumatic rotational subluxation of the axis on the atlas. Rare, predominant in children, of undefined etiology. Late diagnosis leads to neurological sequelae and death. M.E.C.C., female, 5 years old, complaining of neck pain, torticollis and neck stiffness during the past 4 days. She denied having had trauma. History of tonsillitis 15 days before symptom onset, treated with Amoxicillin for 10 days. Computed tomography of the cervical spine showed rotational subluxation of C1 over C2, with diagnosis of type II GS. Conservative treatment was started 6 days after the onset of symptoms, with a Thomas type cervical collar and anti-inflammatory medicine, considering it an acute condition. She was discharged for outpatient follow-up and physical therapy. She had a good clinical outcome after 6 weeks of treatment. At the end of therapy, the patient presented neck alignment without alterations or pain, neck collar removal and medical discharge. GS is a subluxation of the atlanto-axial joint without trauma or bone disease. It usually occurs after head and neck surgeries and upper airway infections. The disease incidence rate and etiology are unknown. Blood dissemination of the septic emboli to the atlanto-axial ligaments is thought to cause joint misalignment. There are four types of atlanto-axial subluxations. In this report, the GS was type II. The diagnosis is clinical-radiological and the gold-standard treatment is conservative for 6 to 12 weeks, including rest, antibiotic therapy, myorelaxants, anti-inflammatory agents, neck traction and neck collar, with good prognosis. Our patient responded well after 6 weeks of treatment, without postural changes or neurological symptoms, with complete symptom remission.

Keywords:
Torticollis,
Cervical Atlas,
Atlanto-Axial Joint.

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INTRODUCTION

Grisel’s syndrome (GS) is defined as the non-traumatic rotational subluxation of the axis (C1) on the atlas (C2). It is a rare syndrome, predominantly affecting the pediatric population. Of unknown etiopathogenesis, there are theories concerning its risk factors: pediatric patients, history of pharyngitis, adenotonsilitis, peritonsillar and neck abscess, otitis media, trauma, upper airway infection, genetic disorders (for example: Down syndrome and Marfan syndrome) and head and neck surgeries. Diagnosis should be established by the association of clinical and radiological findings and should be made as early as possible, with late detection and inadequate treatment causing neurological sequelae and/or permanent facial or neck pain or deformity. The acute form can be treated with rest, antibiotics, anti-inflammatory agents, neck immobilization or traction. Some acute cases and most chronic cases require surgical treatment with skeletal traction or bone fusion.

Despite its rarity, the medical community must be aware of it to avoid underdiagnosis and consequent irreversible injuries.

CASE REPORT

Patient, M.E.C.C., female, 5 years old, admitted to the emergency room, complaining of neck pain, neck deformity and limited range of motion in the past 4 days, (figure 1) using medication to control symptoms, but without improvement. She had no history of trauma. She had bacterial tonsillitis 15 days prior to the onset of these symptoms, and was treated with Amoxicillin for 10 days.

She was referred to the Neurosurgeon and admitted to the hospital for tests. She was submitted to transoral neck radiographs and computed tomography of the neck spine, which showed a rotational subluxation of C1 over C2 (with an anterior displacement of 4.2 mm) (Figure 2) confirming the diagnosis of type II Grisel's syndrome, according to the Fielding-Hawkings classification.

In view of these findings, 6 days after the onset of symptoms, she was treated conservatively with the rigid Thomas-type neck collar (American collar) and received non-steroidal anti-inflammatory drugs, because of the disease’s acute nature. The patient was discharged after 2 days of hospitalization with pain improvement and referred to outpatient follow-up and physical therapy.

The patient had a good clinical outcome, with fixed cervical rotation pattern improvement after 3 weeks of hospital discharge and the neck collar. She was reassessed by the neurosurgeon, who maintained the neck collar for an additional 3 weeks, totaling a period of 6 weeks of treatment. At the end of treatment, the patient presented neck alignment, recovery of the cervical spine range of motion and without pain complaints, neck collar removal and medical discharge.
DISCUSSION

GS is the spontaneous subluxation of the atlantoaxial joint, which usually occurs after an infectious process (48%), and the respiratory tract infection is the most common (83%), followed by retropharyngeal abscess (11%), otitis media (4%) and mumps (2%)⁴, and after surgical procedures in the head and neck region⁴⁻⁵. The incidence rate of the disease and its etiology are unknown, and the currently accepted theory involves the vascular complex that drains the posterosuperior pharynx. This plexus has no lymph nodes, but it has anastomoses between the peri-odontoid venous plexuses and pharyngo-vertebral veins, described by Parker et al., serving as a hematogenous pathway to transport peripheral septic secretions to the superior neck region.

Being close to the transverse, alar and atlanto-axial joint ligaments, it stimulates the inflammatory mediators that cause vascular and synovial engorgement, edema and periligamentous inflammation, thus resulting in a C1-C2 instability. All of this process is associated with greater atlanto-axial ligament laxitude in the pediatric population²⁻⁵⁻⁶⁻⁷.

Corroborating these findings, authors have shown this ligament laxity by the increase in the atlanto-axial interval and engorgement of adjacent neck tissues during infection, using magnetic resonance image⁸. In addition, due to the inflammatory process, C1 and C2 may decalcify, causing a weakening in the transverse ligament insertion⁵⁻⁷⁻⁹, with instability resulting in subluxation with misalignment of the cervical spine, which may evolve with neurological complications.

Although GS is widely seen in children, it can also be found in adults. GS has been reported in patients aged 5 to 12 years and no gender dominance has been reported²⁻³. Of the patients with Grisel’s syndrome, 68% are younger than 12 years⁰. Anatomical differences between these populations may justify this higher prevalence, differences such as: proportionally larger head, greater weakness of the cervical muscles, greater ligament and joint looseness, smaller and more horizontalized C1 and C2 joint faces, undeveloped uncinate process and greater range of motion¹. In this case report, the patient was 5 years old, and the syndrome was installed after a bacterial tonsillitis.

There are four types of atlanto-axial subluxations described by Fielding-Hawkins¹¹. (Table 1) The first type is the C1 rotation over C2 without previous displacement. The second type is the atlas rotation over the lateral reticular process with a 3-5mm anterior displacement. The third type is the atlas rotation with anterior displacement (greater than 5mm) of both atlanto-axial joints; and the fourth type is the atlas rotation with posterior displacement²⁻³⁻¹¹. Subluxation types 1 and 2 are more frequent and neurological symptoms are not common. Spinal cord compression and severe neurological findings can be found in types 3 and 4²⁻⁸. In this case report, the subluxation was type II.

Upon physical examination, three signs reinforce the clinical diagnosis of GS: palpable deviation of the axis’s spinal process in the same direction of the head rotation, sternocleidomastoid muscle spasms ipsilateral to the rotation and antalgic neck stiffness⁸. The palpable deviation of the C2 spinous process in the same direction of the head rotation with atlanto-axial subluxation determines the Sudeck’s sign²⁻³. Patients may present cervicalgia and paresthesia in the upper limbs with neck flexion (Lhermitte’s sign). Pain and limited neck movement characterize the cock-robin position, due to the presence of paradoxical torticollis, which develops by the presence of contralateral sternocleidomastoid muscle (CLSCMM) spasm in the attempt to correct such deformity, different from congenital muscular torticollis, in which the CLSCMM spasticity causes the deformity with the ipsilaterally inclined head and rotation contralateral to the affected CLSCMM²⁻¹². Although less than 15% of the patients show neurological signs and symptoms, extreme consequences such as tetraplegia and sudden death may happen. The torticollis may be an initial symptom of GS, requiring a complete medical history, clinical and radiological examination for diagnostic confirmation.¹³ In our case, the patient had the characteristic Cock-Robin position; however, there were no neurological symptoms.

Radiological evaluation is important for early diagnosis. The atlanto-axial subluxation can be seen by simple transoral x-ray of the cervical spine in search of asymmetry between the C1 lateral masses, due to its rotation, in addition to the apparent wedge shape of this lateral mass in the side skull view radiograph. However, the fixed position of the rotating cervical spine may cause technical difficulties and even mimic a subluxation, rendering such examination unreliable¹⁴. Thus, the gold standard for diagnosis is a three-dimensional computed tomography (CT) of the cranio cervical transition, which allows one to see the C1-C2 rotation, loss of congruence between the joint facets and to calculate the atlanto-odontoid distance (normal distance is ≤ 3mm in adults and ≤ 5mm in children)⁸. CT and magnetic resonance imaging (MRI) are excellent diagnostic tools, and they help classify the

### Table 1. Grise Syndrome types.

<table>
<thead>
<tr>
<th>Type I</th>
<th>C1-C2 rotation without anterior shifting</th>
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<tr>
<td>Type II</td>
<td>C1-C2 rotation with 3-5mm anterior shifting</td>
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<tr>
<td>Type III</td>
<td>C1-C2 rotation with anterior shifting higher than 5 mm</td>
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<tr>
<td>Type IV</td>
<td>C1-C2 rotation with posterior shifting</td>
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GS classification. Nonspecific inflammatory tests are of little value for the diagnosis of this syndrome1,2.

Treatment should be started immediately to prevent progression of the fixed subluxation, prevent neurological complications (paralysis, radiculopathy and death), and cervical ankylosis with permanent limitation of neck mobility, and in chronic cases, plagiocephaly. Conservative treatment is the gold standard and the first line of treatment, especially in acute cases (less than 3 weeks after the onset of symptoms)3. Its indication and execution depend on symptom duration and deformity type according to the aforementioned classification,15 including rest, antimicrobial therapy, muscle relaxants, anti-inflammatory agents, neck traction and soft or rigid neck collar1,2,8,16. If the muscle spasm persists for more than 24 hours, benzodiazepines can be added to the treatment2,3. Still, Wetzel and La Rocca cited by Roncaglio et al. (2016) proposed treatment according to the classification of the subluxation, in which the conservative treatment for type I, II and III lesions would be soft collar, rigid collar (Philadelphia) and halo-vest, respectively, while for type IV lesions, the authors recommend surgical treatment, also indicated for a period of 6 to 12 weeks. For type IV lesions, the authors recommend surgical treatment, also indicated for cases of conservative treatment failure, recurrence of subluxation, and irreducible subluxations12,17. In the present report, consonant with these authors, we chose conservative treatment with the neck collar, which started 6 days after the onset of symptoms (acute phase) and because it was a type II GS, with a positive response and remission of symptoms, after this period of treatment. There was no need for surgery or neck traction.

Prognosis with conservative treatment is excellent when the diagnosis is made in a timely manner, i.e. less than 3 weeks after the first symptoms. After this time, surgical intervention may be required. In most cases, the subluxation reduces spontaneously within 7 days after treatment onset, with the patient recovering neck joint stability8,12. These findings corroborate the findings of Gomes et al. (2011)13, who concluded that Grisel syndrome is a benign disease in most cases, and conservative treatment is sufficient for most patients.

CONCLUSION

GS is a rare syndrome, predominant in children. Early diagnosis and treatment are essential to prevent severe neurological sequelae, including sudden death. Such diagnosis should be made in the presence of torticollis and fixed neck deformities, especially in the pediatric population with a recent history of upper airway infections or head and neck surgeries. The correct diagnosis enables proper GS classification, and it guides the treatment options and the intervention as early as possible. The clinical course is usually benign, especially in the acute forms and, in GS types I and II, which bears good prognosis, the conservative treatment is the first line of treatment. In the present case report, the patient was diagnosed with type II SG, 6 days after the onset of symptoms (acute phase), having undergone conservative treatment and a Thomas type neck collar for 6 weeks, with a good response to therapy with remission of symptoms.

REFERENCES
