CASE STUDY

Grisel’s Syndrome as a Sequela of a Complicated Acute Mastoiditis

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PALABRAS CLAVE
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Abstract  Grisel’s syndrome is a rare complication of ENT area infections. It consists of a non-traumatic atlantoaxial subluxation after an infectious process. Its characteristic symptom is persistent torticollis despite a resolved infection. The knowledge of this condition helps early diagnosis and treatment. We therefore present the case of a patient with Grisel’s syndrome as a sequela of an acute mastoiditis complicated by a Bezold’s abscess.

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Síndrome de Grisel como secuela de una mastoiditis aguda complicada

Resumen  El síndrome de Grisel es una complicación infrecuente de las infecciones que afectan al área otorrinolaringológica. Consiste en una subluxación afaixoidea no traumática, tras un proceso infectioso. Su síntoma característico es la torticolis, dolorosa y persistente, a pesar de haber solucionado la infección. El conocimiento de esta patología ayuda al diagnóstico y tratamiento precoz de la misma. Queremos, por lo tanto, presentar el caso de un paciente con un síndrome de Grisel como secuela de una mastoiditis aguda complicada con un absceso de Bezold, a fin de favorecer el conocimiento de este tipo de proceso.

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Introduction

Grisel’s syndrome is an atraumatic subluxation of the atlantoaxial joint, usually caused by pharyngitis, which is accompanied by oedematous hyperaemia of the transverse and alar ligaments. This leads to greater flexibility of the ligaments, causing a subluxation. When the inflammation disappears, the result is fixation in a rotated position. Pharyngitis may be caused by an infection of the upper


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respiratory tract (tonsillitis, adenoiditis, etc.) or be secondary to surgical interventions in this region (reactive pharyngitis).\textsuperscript{2} It was first described by Sir Charles Bell in 1830, in a patient with a pharyngeal syphilitic ulcer.\textsuperscript{3} In 1930, Pierre Grisel published 2 cases of atlantoaxial subluxation in children.\textsuperscript{4} We report the case of a patient who presented Grisel’s syndrome after acute mastoiditis complicated with a Bezold abscess, in order to increase awareness about this syndrome, given the severe consequences of a delayed diagnosis and treatment of this condition.

Case Report

The patient was a 3-year-old boy with no relevant history except for a dubious allergy to amoxicillin, who presented right otalgia with otorrhea and fever (38.6 C) of 3 days duration despite treatment with oral clarithromycin and topical ciprofloxacin. Subsequently, he presented torticollis and pain in the right mastoid region, so he was transferred to our hospital for evaluation by the Otolaryngology Service. The physical examination found torticollis towards the right side and erythema and pain upon palpation in the mastoid region. The otoscopy found otorrhea originating from a posteroinferior tympanic perforation, with bulging of the posterosuperior wall of the external ear canal. We took a culture of the otic secretion. Upon suspicion of acute mastoiditis, the patient was admitted for intravenous antibiotic therapy (vancomycin 60mg/kg/day and gentamicin 7mg/kg/day). We also performed an urgent computed tomography (CT) scan which showed occupation of the right middle ear and mastoid air cells, with rarefaction of its walls and without coalescence. The patient presented leukocytosis (25,900 mm\(^{-3}\), 70% neutrophils), C-reactive protein (CRP) at 33.52 mg/dl and haemoglobin at 11.7 g/dl. In the culture we identified \textit{Turicella mititidis} resistant to erythromycin and clindamycin. After 5 days of treatment, we observed a worsening of symptoms, as well as anaemia (haemoglobin at 8.4 g/dl) and thrombocytopenia (74,000 mm\(^{-3}\)), leukocytosis (13,200 mm\(^{-3}\)) and CRP at 33.94 mg/dl. We opted for admission to the paediatric ICU due to suspicion of sepsis. We conducted surgical drainage (right atticotomy and mastoidectomy, with exploratory tympanostomy and placement of trans tympanic drainage), and also transfused 2 units of packed red blood cells (haemoglobin at 9.9 g/dl and 387,000 platelets/mm\(^3\)). Seven days later, after clinical improvement, the patient began to complain of severe retroauricular pain, associated with neck stiffness and right otorrhagia. He also presented a fluctuating area in the mastoid tip region. We performed an urgent CT which revealed a Bezold abscess of 2 cm. The collection was surgically drained, leading to clinical and analytical improvement, until the complete resolution of the process. Given the persistence of torticollis during admission despite the resolution of the infectious process, the patient was examined by the Rehabilitation Service. He presented a cervical attitude in left rotation and right lateral flexion, with contracture of the right sternocleidomastoid (SCM) muscle. We began rehabilitation treatment with relaxing massage and cervical mobilisation. Due to the persistence of torticollis, we obtained simple cervical radiographs (anteroposterior, lateral and transbuccal). Upon suspicion of C1–C2 subluxation, we performed an urgent CT scan which confirmed the suspected atraumatic atlantoaxial subluxation (Grisel’s syndrome) (Figs. 1 and 2). We placed a soft, thermoplastic, cervical neck brace. Subsequently, due to the unresolved contracture after intensive rehabilitation treatment, we performed tenotomy of the SCM muscle. Two years after discharge, the patient is currently asymptomatic.

Discussion

Grisel’s syndrome can occur after procedures such as tonsillectomy, adenoidectomy and mastoidectomy. Head and neck infections represent the second most frequent cause.\textsuperscript{5,6}
Sentinel signs are persistent and severe cervical pain, with torticollis and tendency towards head rotation (Table 1). Its pathogenesis is unknown. The most accepted theory explains the onset of the process as a haematogenous spread of infection from the posterosuperior wall of the pharynx towards the cervical spine.7–9 There have been reports of a pathway through pharyngovertebral veins which allows the transmission of infectious emboli up to the periodontoid vascular plexus, without any opposition to such flow. Secondary hyperaemia and abnormal relaxation and bloating of the atloaxoid ligaments lead to a destabilisation of the atlantoaxial joint, with consequent subluxation. The transverse and alar ligaments, which oppose the anterior displacement of the atlas on the axis, appear to be the ligaments involved. Children and patients with Down syndrome, who have an increased laxity of the atloaxoid ligaments and a greater atlanto-odontoid distance, are the most affected. Other authors have proposed that subluxation is secondary and a result of spasms from the cervical muscles when these are exposed to inflammatory mediators transported by a haematogenous route.3

Grisel’s syndrome is usually unilateral. Fielding et al.10 proposed a grading scale to guide its prognosis and therapeutic management. In this scale, type I (the most frequent) is a rotating fixation (not a true subluxation), maintaining stability of the transverse ligament. Most cases improve with muscle relaxants, bed rest, NSAIDs and a soft, stabilising neck brace. In type II is a 3–5 mm displacement of one of the lateral processes of the atlas, requiring cervical traction and reduction with a rigid neck brace and possibly traction with a halo vest for 6–12 weeks. Type III is a bilateral and anterior displacement of over 5 mm and type IV occurs when there is posterior displacement. These last 2 types are more severe and have been associated with spinal cord compression, quadriplegia and even death. They require cervical traction, immobilisation and bed rest. Kraft and Tschopp2 described an additional type 0 (persistent torticollis without radiological evidence of rotating fixation or subluxation). Antibiotic treatment appears to be sufficient in these cases, although intensive monitoring is required to identify a possible progression of symptoms. Surgery with arthrodesis is indicated in cases with insufficient reduction, failure of conservative treatment or recurrent subluxation.

**Figure 2** 3D reconstruction of the CT scan showing a lateral inclination of the atlas on the axis (A: posterior view and B: anterior view) and its rotation, with reference to the transverse processes of the atlas and axis (C: lateral view).

**Table 1** Diagnostic Algorithm of Grisel’s Syndrome.

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<tr>
<th>Feature</th>
<th>Diagnosis</th>
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<tr>
<td>History of ENT surgery or ENT infection.</td>
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<tr>
<td>Torticollis several days after the onset of the infectious process/operation.</td>
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<td>Slight flexion and rotation of the head, with the chin pointing towards the contralateral side.</td>
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<td>Painful active and passive head rotation.</td>
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<td>Elevation of CRP and leukocytes during the first days of torticollis. Subsequent normalisation of these parameters and, in general, absence of fever.</td>
<td>-</td>
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<td>Radiograph of the cervical spine: the space between the atlas and the odontoid of the axis is &gt;5 mm.</td>
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<td>CT: C1–C2 subluxation and rotation.</td>
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CRP: C reactive protein; CT: computed tomography; ENT: ear, nose and throat.

**Conflict of Interests**

The authors have no conflict of interests to declare.

**References**