



## Case Report

# Torticollis in an 8-year-old child due to Grisel's syndrome - A case report

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Received : 01 September 2022

Accepted : 20 September 2022

Published : 04 November 2022

DOI

10.25259/SNI\_805\_2022

Quick Response Code:



## ABSTRACT

**Background:** Grisel's syndrome is a nontraumatic atlantoaxial subluxation resulting from an ongoing local inflammatory process.

**Case Description:** An 8-year-old male presented to the emergency department with neck pain which was sudden in onset without any history of any significant fall or trauma. On physical examination, the patient had torticollis with a head tilt to the left side and the chin rotated to the right. The CT scan confirmed atlantoaxial subluxation with C1 rotated to the right on the odontoid process without anterior displacement. The patient was managed conservatively with antibiotics, anti-inflammatory agents, and head-halter traction.

**Conclusion:** As Grisel's syndrome can have catastrophic outcomes, early diagnosis and treatment are critical. It must be considered in patients presenting with acute torticollis following an infection or prior surgery.

**Keywords:** Atlanto-axial subluxation, Grisel's syndrome, Torticollis

## INTRODUCTION

Grisel's syndrome is a nontraumatic atlantoaxial subluxation resulting from an ongoing local inflammatory process.<sup>[1]</sup> It is more commonly seen in children due to the laxity of the ligaments who present with tenderness of the spinous process and unilateral occipital pain.<sup>[2]</sup> This syndrome is usually attributed to an infection of the upper airways or head and neck regions or with ear, nose, and throat surgical procedures.<sup>[2]</sup> To avoid significant neurological sequelae, it is critical to establish the diagnosis early and initiate prompt treatment. Here, we diagnosed Grisel's syndrome in an 8-year-old child presenting with torticollis.

## CASE PRESENTATION: SHORTEN MARKEDLY

### Clinical and radiographic presentation

An 8-year-old male presented with the sudden onset of neck pain and torticollis characterized by a head tilt to the left side with the chin rotated to the right. He was neurologically intact. Open mouth cervical spine X-ray showed an asymmetric distance of C2 from the lateral masses. The CT

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scan confirmed atlantoaxial subluxation with C1 rotated to the right on the odontoid process in the absence of anterior displacement. The right and left side lateral atlantoaxial joints measured 3 mm and 6.4 mm, respectively. This was diagnosed as a Type I atlantoaxial rotatory subluxation (i.e., Fielding classification). The MRI verified the CT findings and demonstrated that the transverse ligament was intact. However, STIR and T2 images revealed hyperintensity of the pharyngeal and tonsillar tissues and lymph nodes, but there was minimal fluid in the bilateral atlantoaxial joints. The patient was immobilized in a Philadelphia collar and was kept in collar until after all the radiological studies were completed.

#### **Treatment: Antibiotics, anti-inflammatory agents, traction, and bracing**

The patient was put on head-halter traction, 2.5 kg weight was applied. The patient was started on antibiotics and anti-inflammatory agents for a presumed upper respiratory tract infection. Serial X-rays (admission, day 1 and day 2) confirmed further reduction of the torticollis. By day 3, the patient clinically improved and the torticollis had resolved. He was discharged with antibiotics/anti-inflammatory medications for 1 week and wore a Philadelphia collar for 6 weeks. Six weeks later, he was asymptomatic and the X-rays confirmed normal alignment.

#### **DISCUSSION**

Grisel's syndrome is defined as a nontraumatic subluxation of atlantoaxial joint following nasopharyngeal inflammation. It was first described by Sir Charles Bell in 1830 in a patient with syphilis, pharyngitis, and lethal spinal compression.<sup>[4]</sup> Congenital muscular torticollis is the most common form, accounting for 82% of all pediatric patients presenting with torticollis.<sup>[3]</sup> This subluxation likely occurs due to laxity of the ligaments around the atlantoaxial joint due to infection spread hematogenously through the pharyngovertebral veins thus involving the posterosuperior pharynx and periodontoid venous plexus.

#### **Radiological diagnosis and treatment options for Grisel's syndrome**

CT scans with 3D reconstructions are the gold standard for diagnosing Grisel's syndrome. Typically, this should be

supplemented with enhanced MR studies to better define the soft-tissue extent of neural and retropharyngeal pathology.

Early treatment of Grisel's syndrome is straightforward and begins with the administration of antibiotics and anti-inflammatory agents and cervical bracing. If diagnosed later in the clinical course, surgery may be warranted to manage high-grade instability/subluxation.

#### **CONCLUSION**

The diagnosis of Grisel's syndrome must be considered in patients presenting with acute torticollis following either an infection or operative procedure in upper aerodigestive tract. Early diagnosis and treatment are critical to avoid neurological sequelae.

#### **Declaration of patient consent**

Patient's consent not required as patient's identity is not disclosed or compromised.

#### **Financial support and sponsorship**

Nil.

#### **Conflicts of interest**

There are no conflicts of interest.

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**How to cite this article:** Ghodke A, Gadkari A, Trivedi R, Tikoo A, Chaddha R. Torticollis in an 8-year-old child due to Grisel's syndrome - A case report. *Surg Neurol Int* 2022;13:502.