Grisel’s syndrome presenting with neck pain: an atypical case

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ABSTRACT
Grisel’s syndrome is non-traumatic inflammatory subluxation of the atlantoaxial joints presenting clinically as torticollis, neck pain, and reduced neck mobility. Several pathogens have been implicated in its etiology. Early diagnosis and treatment are vital for Grisel’s syndrome to avoid serious neurological complications. This study reports the case of a 6-year-old girl who complained of pain and curvature of the neck following an upper respiratory tract influenza infection. Notably, the patient’s neck pain and curvature worsened toward the end of her normal day for a week. This pattern is rare, but it represents an important example of Grisel’s syndrome as a condition that varies through the day.

Key words: atlantoaxial joints, torticollis, viral infection, pain.

Case Report
A young 6-year-old girl was brought to the Neurosurgery outpatient clinic complaining of pain and curvature of the neck, which she had been experiencing since a week. Her parents explained that the girl’s neck mobility was normal after waking up in the morning, with the curvature and pain in the neck area increasing progressively through the day (Fig. 1). The pain and limited neck mobility were associated with clinical torticollis.

The neck-related problems began following influenza-like symptoms, such as fever and nasal discharge, which started two weeks prior to presentation. Specific blood infection parameters included a sedimentation rate of 38 mm/h, a C-reactive protein level of 74.30 mg/L, and a white blood cell count of 12.6 × 10³ cells/mm³. Reproduction was not observed in any of the body fluid samples sent for culture. Under microscopy, the blood peripheral smear revealed atypical cells (Downey cells), which were considered to have developed secondary to the viral infection.
Three-dimensional (3D) reformatted cervical computed tomography (CT) revealed narrowing at the right anterior of the atlantoaxial joint range on cervical 1 (C1) and cervical 2 (C2), and image matching with subluxation was detected (Fig. 2). Because the patient did not experience torticollis in the early hours of the morning and there was only a small amount of pain at this time, a further 3D reformatted cervical CT scan was performed at 06:00 AM. This revealed that the atlantoaxial distance was normal and that the subluxation had been repaired (Fig. 3).

Superficial neck ultrasonography revealed multiple reactive, ovoid-shaped lymph nodes within the submandibular area on both cervical chains; one on the right was 23 mm long and one on the left was 17 mm long. Contrast-enhanced cervical magnetic resonance imaging (MRI) showed bilateral submandibular reactive lymph nodes and signal variations related to infection inflammation. These exhibited intense contrast and did not form distinct boundaries, and which hold the paravertebral tissues through C1 and C2 vertebrae toward inferior on nasopharynx left lateral wall, have been imaged (Fig. 4).

On the basis of these findings, Grisel’s syndrome was diagnosed and the patient was administered an appropriate antibiotic treatment (ceftriaxone 2 × 50 mg/kg and clindamycin 3 × 40 mg/kg intravenously, and ibuprofen syrup 4 × 20 mg/kg cups orally). Following two weeks of treatment, clinical, laboratory, and radiological findings showed the patient had recovered, and she was discharged home. During the two weeks after discharge, the patient was
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mobilized with a cervical corset. Follow-up examinations at the outpatient clinic one month after discharge detected no pathology and there was no dislocation in the control CT (Fig. 5).

Discussion

Grisel’s syndrome is characterized by rotator subluxation of the atlantoaxial junction of the first and second cervical spines (C1 and C2). It is non-traumatic and rare, appearing after an inflammatory process.\(^1\) \(^3\) Grisel’s syndrome has been reported after otorhinolaryngology infections,\(^4\) \(^6\) or with osseous, ocular, ligamentous, psychiatric, and neurological disorders after head and neck surgery.\(^7\) In addition, torticollis in childhood can be associated with infective agents.\(^8\) The syndrome can be caused by various infectious agents, including *Streptococcus pyogenes*, *Bacteroides ureolyticus*,\(^9\) *Mycobacterium tuberculosis*,\(^10\) *Pseudomonas aeruginosa*, *Staphylococcus aureus*, methicillin-resistant *Staphylococcus epidermidis*, the Epstein–Barr virus, and Kawasaki disease.\(^11\) \(^12\) In our case study, it was concluded that Grisel’s syndrome developed secondary to the patient’s recent upper respiratory tract viral infection.

No microorganisms could be reproduced in the blood or urine cultures, and there were atypical lymphocytes in the peripheral blood smear that supported the occurrence of such a viral infection.

Grisel’s syndrome, which is generally accompanied by torticollis, presents with curvature of the neck, pain in the neck, and limited neck mobility.\(^13\) \(^14\) The torticollis appears very soon after the start of a head and neck infection or following infection of the otorhinolaryngological tracts.\(^15\) Unusually, the patient in our case study did not experience torticollis when she woke in the morning, but the torticollis intensified toward the evening. This pattern has been rarely seen in the literature. In addition, there have been no previous reports of radiology findings that show the absence of subluxation in the morning but its presence later in the day, accompanying the torticollis.

The early diagnosis of Grisel’s syndrome is of vital importance. Serious neurological complications can occur if the condition goes unnoticed or is improperly treated. The condition is diagnosed radiologically using radiographs, CT, and MRI to show atlantoaxial subluxation. It is difficult to diagnose with radiographs alone.\(^16\) The best modality for diagnosing atlantoaxial subluxation is CT, with 3D CT providing the optimal view of this pathology. MRI may show abnormalities in the soft tissue and
nervous tissue that are complementary to the radiological examination.17 These examinations were utilized during the clinical management of the present case.

When Grisel's syndrome is diagnosed early, the primary treatment is conservative. Laboratory tests are required for the early diagnosis and to establish the etiology, and antibiotic treatment, bed rest, external fixation, anti-inflammatory drugs, and muscle relaxants are used against the active infectious agent; plasma exchange or immunoglobulin treatment must be started promptly in cases with a resistant form of infectious agent.18 Treatment selection for the atlantoaxial joint subluxation depends on the severity of the injury. In non-serious injuries, physiotherapy and manipulation may be used; open surgery is indicated for serious subluxations.19 According to the classification of subluxations of the atlantoaxial joint of Fielding and Hawkins20 (Table I), types 1 and 2 represent the most common groups with no neurological deficiency, and types 3 and 4 are the groups that may lead to serious neurological deficiencies associated with spinal cord compression. The subluxations of the atlantoaxial joint were type 1 according to the Fielding classification in our case study.

Grisel’s syndrome should be included in the differential diagnosis for malformation of the occipital, atlas, and axis bones that affect the craniovertebral joint, and foramen magnum malformations. Atlantoaxial instability, infections, trauma, inflammation, malignancy, and Sandifer syndrome21,22 should be ended as atlantoaxial instability like Grisel’s syndrome.

In conclusion, Grisel’s syndrome should be kept in mind for pediatric cases with neck pain and torticollis following a recent upper respiratory tract infection. Painful torticollis that shows a fatal course in the morning and intensifies

Table I. Fielding and hawking classification, reported in 1977.20

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<th>Type</th>
<th>Description</th>
<th>Treatment</th>
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<tr>
<td>Type 1</td>
<td>Most common and is characterized by a simple rotation without anterior displacement of the atlas and the transverse ligament is undamaged</td>
<td>Antibiotics, muscle relaxant, massage therapy, and immobilization with a soft collar</td>
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<tr>
<td>Type 2</td>
<td>The rotatory subluxation is associated with anterior displacement of the atlas ≤ 5 mm and transverse ligament deficit</td>
<td>Reduction and cervical traction with a rigid collar</td>
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<tr>
<td>Type 3</td>
<td>Anterior displacement of the atlas is &gt; 5 mm, both lateral atlantoaxial joints are subluxated anteriorly, the transverse ligament and the articular facets are damaged</td>
<td>Both type 3 and 4 subluxation are highly unstable lesions and, in most cases, associated with neurological symptom. It is necessary cervical traction with “halo vest” and, in the event of neurological symptoms, decompression and arthrodesis of C1 C2</td>
</tr>
<tr>
<td>Type 4</td>
<td>Quite rare, more frequent in adults with rheumatoid arthritis characterized by rotation and posterior dislocation of atlas</td>
<td>Both type 3 and 4 subluxation are highly unstable lesions and, in most cases, associated with neurological symptom. It is necessary cervical traction with “halo vest” and, in the event of neurological symptoms, decompression and arthrodesis of C1 C2</td>
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toward the evening may also indicate Grisel’s syndrome. Progressive neurological deficit and permanent neck deformity can be prevented with appropriate and prompt medical treatment.

REFERENCES


