

Case Report

Retropharyngeal abscess causing grisel's syndrome secondary to tuberculosis of cervical spine, a rare form of torticollis: case report

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ABSTRACT

Grisel's syndrome is a rare type of non-traumatic subluxation of an atlantoaxial joint characterized by torticollis, neck pain, and reduced neck movement. The common causes of Grisel's syndrome are head and neck infection or post-otorhinolaryngology (ORL) procedures. We are reporting a case of a 3-year-old boy with a gradual worsening of neck stiffness, neck pain, and restricted neck movement for more than one month. The patient had no history of trauma. The computerized tomographic (CT) showed a retropharyngeal abscess with a bony erosion causing atlantoaxial subluxation. The management and progress of this patient are discussed. The objective of this case report is to emphasize that Grisel's syndrome should be considered a differential in a painful torticollis to prompt an early diagnosis and treatment to prevent serious neurological complications.

Keywords: Grisel's syndrome, Atlantoaxial subluxation, Retropharyngeal abscess, Tuberculosis

INTRODUCTION

Grisel's syndrome is a rare condition of the cervical spine that refers to a non-traumatic rotatory atlantoaxial subluxation.¹ The main cause of this syndrome is head and neck infections. Post-ORL procedures such as adenoidectomy are also associated with Grisel's syndrome in susceptible children such as those with Down syndrome.² This rare condition predominantly affects children between 5-12 years of age.³ The clinical features are torticollis, neck pain, and reduced in neck movement.⁴ One of the various pathogens that can cause Grisel's syndrome is mycobacterium tuberculosis.⁵ Grisel's syndrome is diagnosed based on the evaluation of clinical and radiological findings.⁶

CASE REPORT

A 3-year-old boy with no significant past medical history presented with gradual worsening neck pain, neck stiffness, and restricted neck movement for more than one

month. The patient had no history of trauma. The physical examination revealed mild torticollis with the head fixed at the right lateral flexion. He also had cervical lymph nodes. The neurological examination was unremarkable with no gross cranial nerve deficits and full strength and sensation in the extremities.

The laboratory investigations showed leucocytosis (1290/mm³), an elevated erythrocyte sedimentation rate (ESR) (40 mm/h), and normal C-reactive protein (<6.0 mg/L). Gastric lavage acid fast bacilli were not detected.

The contrast enhanced tomography (CECT) of the spine demonstrated an increased atlantoaxial distance measuring 1.0 cm and a large hypodense collection at the prevertebral region, suggestive of a retropharyngeal abscess with the bony erosion of C1 and C2, causing atlantoaxial subluxation (Figure 1-2). The contrast enhanced magnetic resonance imaging (MRI) showed a retropharyngeal abscess with a bony erosion of C2 destruction, a superoan-

terior displacement of the dens, and atlantoaxial subluxation.

Parents were counselled for C1/C2 stabilization with instrumentation followed by an intraoral incision and drainage of the abscess; however, they refused surgical intervention and opted for non-invasive treatment. The patient's cervical spine was immobilized with sternal-occipital-mandibular immobilizer (SOMI) orthosis. Based on the radiological finding and clinical presentation, the patient was clinically diagnosed as having tuberculosis (TB) of the cervical spine. The patient was started on an intensive phase of an anti-TB regime (per oral rifampicin 15 mg/kg, isoniazid 10 mg/kg, pyridoxine 5 mg od) and given intravenous (IV) augmentin for 5 days and IV cefazoline for 2 days. The patient was discharged home with a SOMI brace and the continuation of anti-TB treatment for two months. The repeated CECT neck at the two-month post-intensive phase of the anti-TB regime showed a resolving retropharyngeal abscess with bony sclerosis of C1 and C2. The atlanto-axial joint subluxation was persistent. The patient still had limited neck movement with no neurological deficit. The parents still resisted surgical intervention. The patient was then discharged home with a SOMI brace to continue the maintenance phase of the anti-TB regime of 1 year.

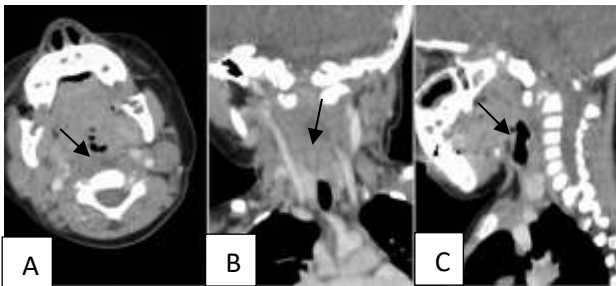


Figure 1: Axial, coronal and sagittal view of contrast enhanced CT shows large hypodense collection at prevertebral region suggestive of retropharyngeal abscess (represented by black arrow).



Figure 2 (A and B): Sagittal and axial view of irregular lytic erosion of the odontoid process and anterior vertebral body of C2 with increased atlantoaxial distance of 1.0 cm (represented by black arrow).

DISCUSSION

Grisel's syndrome is a rare non-traumatic atlantoaxial subluxation joint that occurs following an inflammatory process.^{7,8} This disease is predominant in children, and the usual presentations are torticollis, neck pain, and reduced neck movement.⁹

The main aetiologies of this rare condition are head and neck infections, such as tonsillitis, pharyngitis, tonsillar abscess, or cervical abscess, or post-otolaryngology procedures, such as tonsillectomy, adenoidectomy, and repair of the choana atresia.⁹

There are theories that explain the pathophysiology of Grisel's syndrome; however, the exact pathogenesis still remains unclear and continues to be debated.¹⁰ One of the theories is the two-hit hypothesis, which is proposed by Battiatà and Pazos (2004).

The "first" hit explains the atlantoaxial subluxation, which occurs from a pre-existing cervical laxity that is seen in children. The second "hit" of the hypothesis describes the event of a cervical muscle spasm that is caused by inflammatory mediators, which are transported from the retropharyngeal space into the soft tissue of the atlanto-axial space through the pharyngovertebral plexus during head and neck infections.¹⁰

The gold standard to establish the diagnosis of Grisel's syndrome is based on the radiological finding of a CT scan of the neck. Subluxation is detected when there is an increase in the atlanto-axial interval of more than 4.5mm in children.¹⁰ In the presented case, the CECT demonstrated C1 and C2 subluxation in the presence of a retropharyngeal abscess.

Grisel's syndrome that is detected early is treated conservatively. Treatment includes complete bed rest, neck immobilization with orthosis, antibiotic therapy, muscle relaxants, and anti-inflammatory therapy.¹¹ This is because early conservative intervention prevents permanent neck deformity.¹¹

However, delayed diagnosis after three weeks of initial symptoms leads to a greater risk of recurrence and a permanent neck deformity that requires surgical therapy. This is because the alar and transverse ligament undergo chronic changes.¹²

Fielding and Hawkins classification describes the atlantoaxial subluxation based on the axial CT scan (Table 1).¹³ Type I and II are the most common subluxations with no neurological deficit, whereas type III and IV are rare and have the potential to lead to neurological deficits associated with spinal cord compression.

Table 1: Fielding and Hawkins classification of Grisel's syndrome.¹³

| Staging | Atlantodental interval (ADI) |
|---------------|---|
| Type 1 | Fixed rotation of atlas and axis without anterior displacement (atlanto-axial interval <3 mm) |
| Type 2 | Rotatory fixation with anterior displacement of the atlas of 3-5 mm |
| Type 3 | Rotatory fixation with anterior displacement of the atlas of >5 mm |
| Type 4 | Rotatory fixation with posterior displacement of the atlas |

The management for type I is mainly conservative with antibiotics and a soft cervical collar, whereas for type II subluxation, reduction and cervical traction is needed with a rigid collar.

Fielding's type III and IV subluxations are unstable and may be associated with neurological deficits. Thus, the definite management is cervical traction with a halo vest and the arthrodesis of C1-C2 in the events of neurological deficits.¹³

Based on Fielding and Hawkins classification, our present case was consistent with Type III subluxation. Hence, the definite treatment is surgical intervention, which is C1/C2 stabilization with instrumentation followed by an intraoral incision and drainage of the retropharyngeal abscess.

Nevertheless, in our case, the atlantoaxial subluxation was persistent with C1 C2 bony sclerosis, which confirms that the delay in definitive treatment caused chronic changes in the transverse and alar ligaments, leading to a permanent neck deformity.

In our case, tuberculosis (Tb) of the cervical spine is diagnosed based on multiple factors. One is based on the radiological finding of the CT neck, which demonstrated destruction of C2 vertebra with an abscess collection. These are significant radiological features of a Tb spine.^{14,15}

Sabah is a geographic region that carries a high burden of the Tb rate. It is reported that Sabah accounted for 20% of Malaysia's TB cases despite representing only 10% of Malaysia's population. Therefore, the diagnosis of tuberculosis of the cervical spine should be considered in regions such as Sabah.¹⁶

Moreover, the radiological evidence of a resolving retropharyngeal abscess in this case shows that the abscess responded well to anti-Tb treatment. According to the Malaysian Tuberculosis (Tb) guidelines, the regime for children with a Tb spine is 2 months of an intensive phase with Isoniazid, Pyrazinamide, Ethambutol, and Rifampicin and another 10 months of the maintenance phase of Isoniazid and Rifampicin.¹⁷

CONCLUSION

Grisel's syndrome is a rare condition but an important differential in paediatric cases with painful torticollis. Early and effective treatment leads to a good prognosis. The present case underscores the importance of an early diagnosis and treatment of Grisel's syndrome to prevent permanent neck deformities and serious neurological deficits.

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