



CASE REPORT ON GRISEL SYNDROME: AN OVERLOOKED CAUSE OF ACUTE TORTICOLLIS IN A CHILD

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ABSTRACT

Introduction: Torticollis is a common clinical presentation in a child which can occur due to multiple underlying etiologies. Grisel syndrome remains a rare entity to be diagnosed early in a child presenting with acute atraumatic torticollis. **Case Presentation:** We report such a case of retropharyngeal abscess in a 7-year-old boy who presented with acute torticollis and associated atlanto-axial subluxation. This case is unique in the way it was timely diagnosed with appropriate imaging, where muscle spasm was kept as the most plausible clinical cause of the presenting complaints. Patient was timely started on broad-spectrum antibiotics with resultant resolution of symptoms and complete restoration of normal neck movements. **Conclusion:** Grisel syndrome is an uncommon complication of upper respiratory tract and neck infections. A comprehensive history followed by multimodality imaging is indicated for early diagnosis and treatment. An awareness of this entity amongst physicians and radiologists is important for preventing development of irreversible torticollis.

KEYWORDS : Grisel syndrome - Torticollis - Atlantoaxial rotatory subluxation - Retropharyngeal abscess - cervical lymphadenitis

INTRODUCTION:

Torticollis or wry neck is a clinical finding characterised by rotational deformity of cervical spine with secondary head tilt. It can be a congenital or acquired deformity. Muscle spasm is the most common cause of acquired acute onset atraumatic torticollis in children. Non muscular causes have rarely been attributed as the underlying etiology. However, sometimes non-muscular etiology with associated muscle spasm can also result in painful torticollis. One of the causes is atlanto axial rotatory fixation which is a rare but important diagnosis. It is the rotational disorder of atlanto axial joint which leads to limited rotation or (in rare cases) fixation of the neck. The term Grisel syndrome refers to atlanto axial rotatory subluxation secondary to infectious or inflammatory processes of the head and neck region, such as sinusitis, otomastoiditis, pharyngitis or retropharyngeal abscess.

CASE REPORT:

A 7-year-old boy presented to paediatrics OPD with complaints of painful neck movements with recent onset torticollis. There was no history of trauma or pre-existing health condition. History of intermittent episodes of fever for 3 days was associated with presenting complaints. On examination, the patient's head was rotated to right and chin tilted to left suggesting right-sided torticollis {Figure 1(A)} that was painful on any attempted neck movement. There was limited range of motion of the neck secondary to pain. The boy showed mild motor weakness in right upper limb with reduced power compared to contralateral side. Multiple cervical lymph nodes were palpable.

Clinical differentials of muscle spasm and cervical lymphadenitis were considered.

Patient was further advised an ultrasonography of neck to confirm the clinical diagnosis of acute lymphadenitis which showed multiple ovoid enlarged reactive cervical lymph nodes at bilateral cervical stations II, III, IV and V, largest on right side. None of the lymph nodes showed necrotic changes. Bilateral sternocleidomastoid muscles appear normal and symmetrical in echogenicity and bulk with no soft tissue

lesion. Also radiographs soft tissue neck were performed. Lateral radiograph of neck {Figure 1(B)} showed loss of normal cervical lordosis with mildly increased width of the prevertebral soft tissue anterior to the cervical spine with no significant airway compromise. CT scan cervical spine {Figure 2} showed atlanto-axial dislocation with asymmetric odontoid-lateral mass interval (right > left). Visualised vertebral bodies appear normal with no e/o bony erosions/destructions.

MRI neck {Figure 3} revealed a well-defined elongated collection in the retropharyngeal space extending from the level of atlas to first dorsal vertebra in its cranio-caudal extent suggesting retropharyngeal abscess with bilateral cervical lymphadenopathy. The collection at its superior aspect showed extension into the space between dens and right lateral mass of atlas and inferiorly showed an extension posterior to right carotid space into superior mediastinum with resultant anterior displacement of right carotid vessels. Also, multiple discrete and coalesced lymph nodes were seen at bilateral cervical and right paratracheal stations.

In this case, cause of torticollis was likely retropharyngeal abscess associated with cervical lymphadenitis resulting in associated atlantoaxial rotatory subluxation (Grisel syndrome). Patient was kept on broad spectrum antibiotics and to which he responded well with restoration of normal neck movements and complete resolution of neck tilt on six weeks follow up.



Figure 1 (A) Clinical image of child with head tilt to left (B) Lateral radiograph shows increased Prevertebral soft tissue shadow at the upper cervical spine

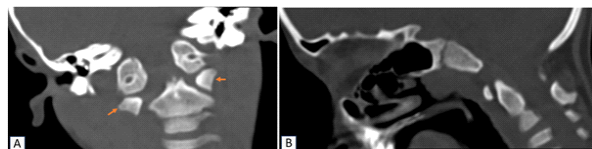


Figure 2 (A) CT coronal section of cervical spine with asymmetric odontoid-lateral mass interval (more on right than left) (arrow points at lateral mass of atlas on each side) (B) Sagittal CT of cervical spine shows normal anterior atlanto-dens interval (< 3 mm)

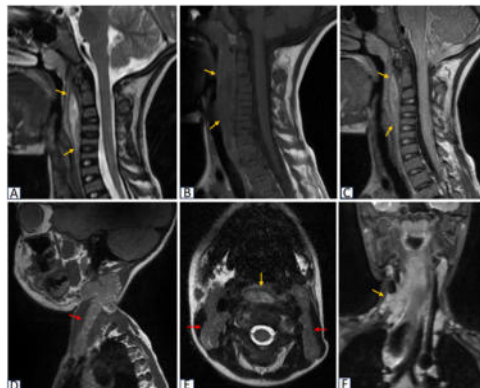


Figure 3 (A, B, C) Sagittal T2, T1, PD images show retropharyngeal abscess (yellow arrow) causing reversal of cervical lordosis and mild narrowing of airway. (D) Parasagittal T2WI shows enlarged cervical lymph nodes (red arrow) on right. (E) Axial T2WI shows retropharyngeal abscess (yellow arrow) with enlarged bilateral cervical lymph nodes (red arrows). (F) Coronal T2WI shows retropharyngeal collection extending on right in superior mediastinum

DISCUSSION:

Torticollis is a congenital or acquired deformity characterized by rotational deformity of the cervical spine with secondary tilting of the head. Congenital torticollis can be caused by different underlying pathologies of muscle (eg: congenital muscular torticollis, fibromatosis coli), skin (pterygium coli), bone (segmentation anomalies), central nervous system (syringomyelia), ligamentous laxity (down syndrome, osteogenesis imperfecta).

Acquired torticollis, seen in older children and adolescents, is often secondary to trauma with infection, or tumours being less common causes [2].

In cases of atraumatic acquired torticollis, Grisel syndrome is rare but also an overlooked entity. It can be associated with upper airway infection, neck infection or inflammation [3]. The higher predilection in children is considered to be a result of multiple factors which include : hypermobile C1 on C2, greater ligamentous laxity in the cervical region, soft bones, horizontally oriented facet joints, and a higher rate of upper airway infections [4]. Initially Grisel thought of muscle spasm to be the cause of subluxation however, now another hypothesis of hyperemia following infection resulting in decalcification of anterior arch of C1 and laxity of transverse ligament is also considered as an important cause behind the subluxation [5]. Also, inflammatory mediators reaching atlanto-axial joint via periodontoid vascular plexus may cause synovial and vascular congestion, periligamentous inflammation, and oedema contributing to subluxation [6].

Fielding and Hawkins classified subluxation into 4 types based on severity [7] :

Type I: Rotatory atlantoaxial dislocation without anterior displacement of the atlas (i.e. atlantodental interval < 3 mm)

Type II: Rotatory atlantoaxial dislocation with anterior displacement of the atlas by 3–5 mm

Type III: Rotatory atlantoaxial dislocation with anterior displacement of the atlas > 5 mm

Type IV: Rotatory atlantoaxial dislocation with posterior displacement of the atlas

The primary treatment for Grisel syndrome is conservative/medical if diagnosed timely [8].

Following meticulous history and correlating findings with clinical examination, torticollis should be classified under congenital or acquired category, followed by appropriate imaging modality depending on list of possible clinical differentials as was done in our case. Ultrasonography is the first modality for suspected congenital torticollis in neonates and infants. In cases of acquired torticollis further differentiation into traumatic or atraumatic can be made depending on clinical history. Cross sectional imaging CT/MRI should be performed to understand the underlying etiology. Dynamic CT with 3D reconstruction is the best imaging modality for classifying atlanto-axial rotatory dislocation appropriately [2]. However MRI helps in detailed evaluation of soft tissue and ligaments with early identification of inflammation and subluxation if meticulously evaluated.

Early diagnoses of Grisel syndrome can only be made in the presence of awareness and familiarity of radiologist with this entity followed by required imaging evaluation. Timely diagnosis is crucial to prevent long term neurological complications and permanent neck deformities.

CONCLUSION:

This case report underscores the importance of considering the rare entities like Grisel syndrome in the differential diagnosis of acute onset atraumatic torticollis. Grisel syndrome remains an uncommon complication of upper respiratory tract and neck infections. A comprehensive history followed by appropriate use of ultrasonography, radiography and cross-sectional imaging modality is indicated for early diagnosis and treatment for improving patient prognosis. Both physician and radiologist should be aware of this rare, dangerous yet potentially treatable entity in case of acute atraumatic torticollis.

KEY MESSAGES:

- Acute onset atraumatic torticollis needs early diagnosis of underlying etiology and appropriate timely treatment.
- Grisel syndrome is a rare but known cause and complication associated with torticollis which is often overlooked.
- Early identification of rare entity of Grisel syndrome is only possible with the help of adequate imaging investigation involving cross sectional imaging modalities, which is crucial for prevention of long term neurological complications and permanent neck deformities.

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