Pearls & Oy-sters: Grisel Syndrome Presenting as Pseudodystonia: A Twist in the Neck

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Abstract

Pseudodystonia is a term that describes abnormal postures, repetitive movements, or both, where clinical analysis, imaging, laboratory, or electrophysiologic investigations indicate that these movements are not consistent with dystonia. Grisel syndrome (GS), characterized by rotatory subluxation of the atlantoaxial joint (AAJ) due to nontraumatic causes, is a cause of pseudodystonia. GS is seen in children less than 12 years of age and should be suspected in patients with acute onset of painful torticollis. We report 2 girls, aged 9 and 6 years, who developed painful torticollis following upper respiratory tract infection. They were thought to have cervical dystonia and referred to a movement disorder specialist for botulinum neurotoxin therapy (BoNT). MRI of the cervical spine showed type I and type II rotary AAJ subluxation, respectively, which confirmed the diagnosis of GS. Short tau inversion recovery hyperintensity was noted suggesting AAJ edema without any bone erosion or cord compression. Abruptness of onset, presence of severe pain, resistance to passive neck movements, fixed postures present equally in rest and action, absence of sensory trick, and persistence in sleep favor pseudodystonia. Both subjects improved with conservative treatment, which included temporary immobilization of the cervical spine and anti-inflammatory drugs. Early identification and treatment is imperative to avoid neurologic complications, like high cervical compressive myelopathy, which can lead to quadriplegia and respiratory distress. Prominent sternocleidomastoid contraction ipsilateral to the rotated chin helps to clinically identify GS.

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Video

Pearls

- Pseudodystonia is a term that describes abnormal postures, repetitive movements, or both, where clinical analysis, imaging, laboratory, or electrophysiologic investigations indicate that these movements are not consistent with dystonia.
- GS is characterized by rotatory subluxation of the AAJ due to nontraumatic causes and is a cause of pseudodystonia.
- GS is seen in children less than 12 years of age and should be suspected in acute-onset painful torticollis.
- Early identification, conservative treatment with immobilization of the cervical spine, and anti-inflammatory drugs are imperative to avoid neurologic complications.

Oy-sters

- GS may be mistaken for cervical dystonia; patients may be erroneously referred to a movement disorder specialist for BoNT.
- Prominent sternocleidomastoid (SCM) contraction ipsilateral to the rotated chin associated with restriction of passive neck movements helps to clinically identify GS.

Case 1

A 9-year-old girl presented with a 2-month history of head tilt toward the right, which was first noticed acutely when she awoke. The head tilt was initially intermittent and then progressed to sustained posturing throughout the day. It was persistent in sleep. The patient reported pain and restricted neck movements. Chewing and movements of the jaw initially caused pain. There

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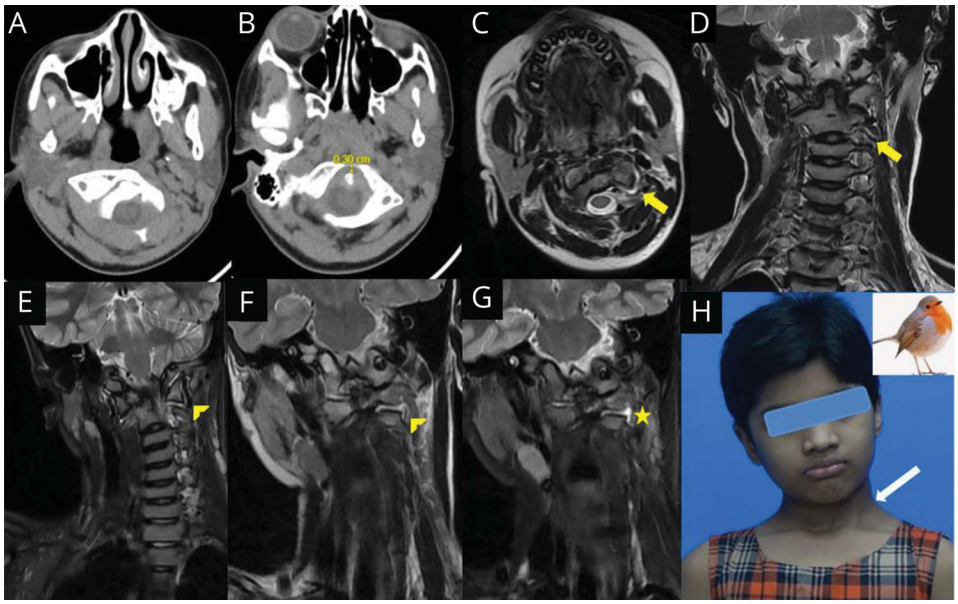
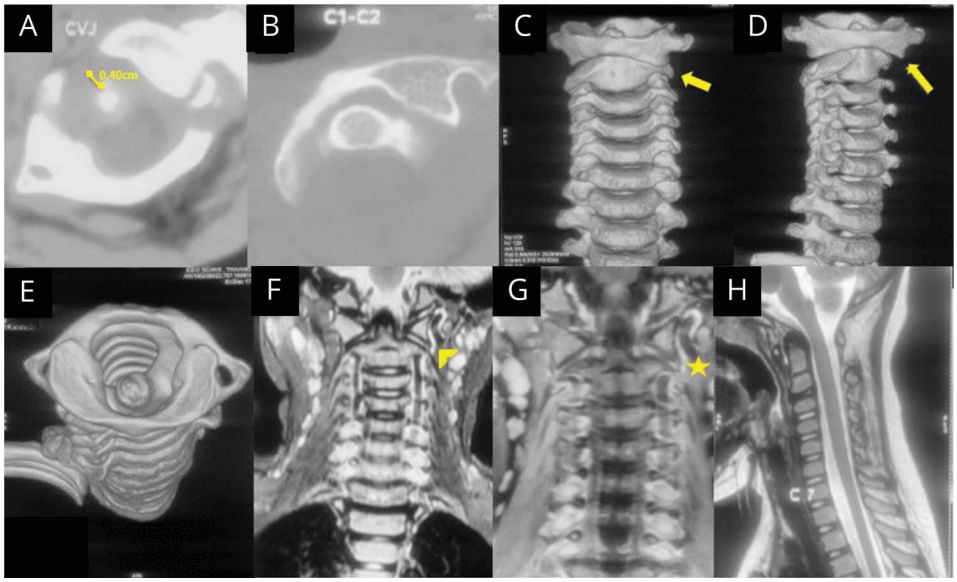
Figure 1 Type I Atlantoaxial Rotary Subluxation in Case 1



Cervical spine CT axial images (A and B) show atlantoaxial joint (AAJ) type I subluxation with mild anterior displacement (≤3 mm) of body of atlas over the axis. MRI of the cervical spine T2 axial (C) shows no evidence of cord compression, and coronal (D) images corroborate the AAJ subluxation with increased space between left lateral mass of C1-C2 (arrow). Short tau inversion recovery (STIR) (E–G) shows moderate effusion of the left AAJ with hyperintensity in the subarticular surface of left lateral mass (arrowheads) and edema (star) of left AAJ synovia and adjacent paraspinal soft tissue without any bone erosion. Characteristic cock-robin head tilt posture seen in patients with Grisel syndrome (H) with prominence of ipsilateral sternocleidomastoid muscle (white arrow); inset shows cock-robin head posture which patients with GS resemble.

were no abnormal movements elsewhere in the body. She had an upper respiratory tract (URT) infection before the onset of symptoms, but denied fever, illicit drug ingestion, or trauma. She was given a diagnosis of cervical dystonia and initiated on trihexyphenidyl and referred to our facility for botulinum neurotoxin therapy (BoNT). Examination revealed a bright child (height 135 cm, 50th centile), with torticollis and chin rotation to the left, spasm and prominence of the left SCM, and painful restriction of neck movements (Video 1, segment 1). There was no pharyngeal congestion or palpable lymphadenopathy. Neurologic examination was otherwise normal. Investigations showed normal white cell count, C-reactive protein, erythrocyte sedimentation rate, and a negative vasculitis profile. MRI of the cervical spine showed type I rotary atlantoaxial joint (AAJ) subluxation with moderate effusion and increased space between the left lateral mass of C1-C2 with subtle anterior displacement (<3 mm) of body of atlas. Short tau inversion recovery hyperintensity was noted in the subarticular surface of the lateral mass with edema of adjacent left paraspinal soft tissue without bone erosion (Figure 1, A–G). She was

Figure 2 Type II Atlantoaxial Rotary Subluxation in Case 2



CT of the cervical spine (A and B) shows type II atlantoaxial rotary subluxation as illustrated by the CT reconstruction images (C–E, arrow). MRI cervical spine T2 image (F) shows left AAJ hyperintensity (arrowhead) and mild contrast enhancement (star) in T1 postcontrast image (G) suggesting effusion. There was no evidence of cord compression (H).

initiated on analgesics, short course of antibiotics, anti-inflammatory agents (i.e., hydroxychloroquine, sulfasalazine, and short-course oral corticosteroids), and Philadelphia collar for neck immobilization. Over the next 3 weeks, she became asymptomatic with complete recovery of her torticollis and neck movements (Video 1, segment 2).

Case 2

A 6-year-old girl with a 2-week history of acute-onset fixed painful posturing of the neck with leftward head tilt, 1 month following a URT infection, was referred for BoNT. Examination revealed torticollis with chin rotation toward the right, prominence of the right SCM, and restriction of all neck movements (video 1, segment 3). The rest of the examination was normal. Blood parameters were within normal limits. X ray of the cervical spine showed subluxation of the AAJ, which was confirmed by CT showing type II atlantoaxial rotary fixation (anterior displacement of atlas by 3–5 mm) (Figure 2, A–E). MRI of the cervical spine showed T2 hyperintensity of the AAJ with mild contrast enhancement suggesting effusion without cord compression (Figure 2, F–H). She was treated with analgesics, anti-inflammatory agents, and Philadelphia collar for neck immobilization, and she had a complete recovery in 14 days (Video 1, segment 4).

Discussion

We present 2 children with Grisel syndrome (GS), initially misidentified as cervical dystonia, who improved completely with accurate diagnosis and conservative treatment. Pseudodystonia is defined as abnormal postures and/or repetitive movements resulting from causes not compatible with dystonia.[1] GS is a clinical condition characterized by a rotatory subluxation of the AAJ due to nontraumatic causes. Although rare, it is a potentially fatal cause of pseudodystonia in children below 12 years of age. GS should be suspected in children with acute-onset torticollis, restricted painful neck movements, and prominent SCM ipsilateral to the rotated chin, especially in the setting of a recent URT infection or intervention (e.g., tonsillectomy, adenoidectomy, mastoidectomy, tympanoplasty, uvulectomy, or cochlear implantation). Early identification and treatment is imperative to avoid complications like high cervical compressive myelopathy.[2,3]

Identification of pseudodystonia is critical in formulating a diagnostic and treatment approach. Pseudodystonia causing torticollis can be due to various etiologies. They can be classified into musculoskeletal disorders without coexisting neurologic abnormalities (GS, congenital vertebral anomalies such as Klippel-Feil syndrome, congenital muscular torticollis [a rare congenital musculoskeletal disorder with unilateral shortening of SCM]), lesions of the posterior fossa and cervical cord (tumors/cysts), primary neurologic conditions (trochlear/abducens palsy and vestibulopathy), benign paroxysmal torticollis, infections like retropharyngeal abscess, and Sandifer syndrome (SS).[1,4] In SS, it is thought that the tilted head position probably yields relief from the discomfort caused by acid reflux. Abrupt onset, severe pain, resistance to passive neck movements, fixed posture present equally at rest and action, absence of sensory trick, and persistence in sleep favor pseudodystonia.[1] Patients with dystonia have insidious onset (except due to dopaminergic blocking agents and rapid onset dystonia-parkinsonism), experience sleep benefit, lack painful restriction of joint movements, and have action-induced overflow dystonia to unaffected muscles initiated by voluntary movement overtime. Antecedent URT infection or surgery is an additional clue to GS.[2] Severe pain is also associated with other pseudodystonias, such as stiff-person syndrome or painful tonic spasms of multiple sclerosis and neuromyelitis optica.[1]

Initially described by Sir Charles Bell in 1830, GS was named after a French otorhinolaryngologist, Pierre Grisel, who reported 3 such children in 1930.[3] Although most cases were reported following URT infection (57.7%) and otorhinolaryngeal surgeries (37.8%), there have been reports of GS following gastrointestinal infections, meningitis, and Kawasaki disease (KD) with a mean delay in diagnosis being 33 days.[2] Two-thirds of cases are in children, without any sex predilection. Greater predominance in children is ascribed to considerable ligamentous laxity of the cervical region, hypermobility of C1 on C2 with greater atlas-dens interval, weak cervical muscles, immature bone formation, horizontally oriented facet joints, larger synovial folds in occipito-AAJs, and an increased frequency of URT infections.[2] Alar and transverse ligaments are characteristically lax in children. A system of pharyngovertebral veins which originate in the posterosuperior pharynx, traverse the prevertebral fascia, and drain in the periodontoidal plexus. This extends inflammation from the pharynx to the atlantoaxial ligaments, leading to ligamental laxity aggravated by regional hyperemia and inflammation.[3] Association with diseases harboring ligamentous laxity like Down and Marfan syndrome has led to the currently postulated 2-hit hypothesis of GS, implicating muscle spasm following an infection or surgery in a child who is predisposed by lax cervical ligaments.[2]

The association of GS with KD has led to the postulation of AAJ subluxation due to postinfectious immune-mediated joint inflammation similar to Reiter syndrome. GS can occur as a complication in 0.6% patients with KD, all of whom had cervical lymphadenopathy and received IV immunoglobulin with aspirin within 10 days from fever onset along with cervical collar and resolved without neurologic impairment.[5]

This acute painful torticollis has been labeled as cock-robin position [Figure 1 H (inset)], and children find it impossible to turn the head past the midline even transiently. Prominence of the SCM ipsilateral to the tilted chin, contrary to the prominent contralateral SCM in cervical dystonia, is highly suggestive of GS.[2,3] This is a compensatory muscle spasm to limit the head tilt, induced by joint subluxation. Dystonic torticollis is a basal ganglia disorder induced by aberrant

afferent inputs from the cerebellum characterized by loss of inhibition within the sensorimotor circuitry, disrupted sensorimotor integration, and maladaptive homeostatic plasticity where the bulkier, overactive SCM is contralateral to the side of tilted chin.

MRI of the cervical spine would be the first investigation for making a diagnosis and ruling out other causes of pseudodystonia by evaluation of cervical soft tissues, ligaments, and spinal cord. CT of the cervical spine with 3-dimensional reconstruction is the gold standard for confirmation and grading of AAJ subluxation. The Fielding Hawkins classification grades the rotary subluxation of the AAJ as type I (rotation of the atlas and ≤3-mm anterior displacement), type II (3–5-mm displacement), type III (>5-mm displacement), and type IV with posterior displacement, of which the last is very rare.[6] Type I and II (our case scenarios) are more common and are amenable to conservative management with antibiotics, anti-inflammatory drugs, and external immobilization, whereas type III and IV may need more aggressive surgical management.[7] Literature review shows that only 12% of GS had undergone surgery as a first- or second-line treatment.[2] Complications in GS may vary from local pain, paresthesia, lower cranial nerve palsies, quadriparesis/plegia, and death.[2,3] Identifying the clinical clues in an acute-onset painful torticollis in a child and confirmation of GS with CT or MRI are cardinal in management of this treatable condition.

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Disclosure

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Appendix Authors

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Divya K P, MD, DM	Department of Neurology, Sree Chitra Tirunal Institute for Medical Sciences and Technology, Thiruvananthapuram, Kerala, India	Conception, organization, and execution of the research project; writing of the first draft; and the review and critique of the manuscript
Ajith Cherian, MD, DM	Department of Neurology, Sree Chitra Tirunal Institute for Medical Sciences and Technology, Thiruvananthapuram, Kerala, India	Conception, organization, and execution of the research project and review and critique of the manuscript

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