

CASE REPORT

Inflammatory C2–3 subluxation: a Grisel's syndrome variant

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The concurrence of non-traumatic atlanto-axial subluxation with inflammation of the adjacent neck tissues is known as Grisel's syndrome. We report a 5 year old boy with recurring episodes of head tilt and painful and restricted neck movements that developed after repeated bouts of sinusitis. Radiographs showed a subluxation of the C2–3 joint. Medical treatment, with cervical collar, physiotherapy, and non-steroid anti-inflammatory agents, led to complete cure of the disease. We suggest that Grisel's syndrome can occur in a location different from the classic atlanto-axial joint. To the best of our knowledge, this is the first report of a symptomatic case of Grisel's syndrome occurring at the C2–3 segment.

Atlanto-axial rotatory subluxation has been reported in a variety of conditions, namely trauma, infection, osseous and ligamentous diseases, and after ENT surgical procedures.^{1–3} Grisel's syndrome refers to a post-infectious dislocation of the atlanto-axial joint that produces clinical manifestations, namely head tilt and restricted and painful neck movements.^{1–3} The entity is seen almost exclusively in children. Lopes and Li recently reported two asymptomatic patients with the incidental finding of a C3–4 subluxation of post-infectious nature and suggested this condition was a variant of Grisel's syndrome.⁴ We report a child who presented with clinical manifestations of Grisel's syndrome but who had a C2–3 subluxation, showing that Grisel's syndrome can occur in a different place from the classically described atlanto-axial location. To our knowledge this is the first report in the literature of a symptomatic variant of Grisel's syndrome occurring at the C2–3 region.

CASE REPORT

A 5 year old boy was referred to our unit for assessment of recurring episodes of painful torticollis. Family and past histories were unremarkable; there was no traumatic antecedent. Since the age of 3 years, the child had suffered repeated bouts of sinusitis. During the previous year he had experienced three episodes of torticollis and painful cervical movements that closely followed each attack of sinusitis. Cervical symptoms in each episode lasted about two weeks. He had neither headaches nor diplopia. His neurological examination was normal, except for a head tilt and restricted and painful neck movements. Cervical spine radiographs showed a tilted head posture with a rotational component in the anteroposterior view (fig 1) and loss of the normal alignment of the spinolaminar line in the lateral projection (fig 2). Lateral flexion-extension radiographs of the child's cervical spine showed that the subluxation did not modify in extension but that it markedly reduced in flexion. The child was managed conservatively for six weeks with mild physiotherapy, a soft



Figure 1 Anteroposterior cervical spine radiograph of the child showing a slight head tilt and rotation of the spinous processes of C2 and C3 (arrows).

collar, and non-steroidal anti-inflammatory agents with success. There have been no recurrences after one year follow up.

DISCUSSION

Grisel's syndrome represents a subluxation of the atlanto-axial joint not associated with trauma or bone diseases and is primarily a disorder of childhood.^{1–3} Infrequently reported in the neurosurgical literature, the syndrome is apt to occur in association with any condition that results in hyperaemia and pathological relaxation of the transverse ligament of the atlanto-axial joint.^{3–5} It has been described in common otolaryngic entities such as tonsillitis, pharyngitis, otitis media, adenoiditis, parotitis, and tonsillar or cervical abscesses, or after common surgical procedures such as tonsillectomy, adenoidectomy, or mastoidectomy.^{2,3,5,6} The condition is thought to be caused by the spread of septic exudates through venous and lymphatic channels that connect the peripharyngeal space and the cervical venous plexus.³ Nasopharyngeal inflammation causes hyperaemia that may weaken the transverse and alar ligaments and the articular capsules resulting in atlantoaxial instability.^{3,5} Characteristically, the syndrome has almost exclusively been reported in children, although infrequent adult cases have also been documented.⁷ The predominance of the syndrome in children and adolescents may be explained by the hypertrophic status of the peripharyngeal lymphoid tissue in the first years of life. The



Figure 2 Lateral radiograph of the child's cervical spine. Note the extent of the retrolisthesis C2-C3 (lines).

adenoids and pharyngeal lymphoid ring, with their childhood predilection for acute and chronic inflammation, are situated in the region drained by the pharyngovertebral plexus.

The classic location of Grisel's syndrome is at the atlanto-axial joint.^{2,3,6} The diagnosis of Grisel's syndrome is based on the association of clinical findings (head tilt with painful and restricted neck movements) with a previous history of an upper respiratory tract infection.^{1,3} Radiographs and computed tomography scans of the cervical spine establish the diagnosis. Early management—consisting of cervical immobilisation, medical treatment, and physiotherapy—is considered the key factor for achieving a satisfactory outcome.^{2,3,6} Cases treated inappropriately may result in a fixed and painful neck deformity that may even require surgical fusion.⁷ To our knowledge, the only affected cervical segment thus far reported in Grisel's syndrome is the atlanto-axial joint.^{1-3,5,7} Coexisting atlanto-axial and atlanto-occipital subluxation from neck infection were described by Hettiaratchy and colleagues.⁸ Interestingly, Lopes and Li have recently documented two children with midcervical (C3–4) ligamentous instability and labelled the disorder "a variant of Grisel's syndrome".⁴ These authors suggested that the pathogenesis of this condition was similar to that of Grisel's syndrome.⁴ However, their two patients were asymptomatic with regard to the cervical subluxation; the disorder was an incidental finding in the radiographs obtained to evaluate the peripharyngeal soft tissues.⁴ We hypothesise that the mechanism of the C2–3 subluxation in our patient is probably identical to that of Grisel's syndrome. The vertebral displacement was caused by inflammation related hyperaemia together with abnormal laxity of the ligaments. The slippage is favoured by the marked hypermobility between the second and third cervical vertebrae seen in children, which is caused by the horizontal disposition of the articular processes of the upper joints of the cervical spine in children. Another peculiarity of

our patient is the direction of the dislocation. Lopes and Li's patients exhibited a C3–4 anterolisthesis, while our patient showed a C2–3 retrolisthesis.

Differential diagnosis in our patient was made against cervical bone anomalies, tumours of the posterior fossa and spinal cord, cervical spine trauma, ocular and vestibular disorders, and dystonic torticollis associated with the untoward effects of drugs. We also took into account the peculiarities in mobility of the cervical spine of children, namely the physiological pseudosubluxation of the infantile cervical vertebrae.² In this regard, a reliable sign consists of the misalignment of the involved vertebrae, which was assessed by drawing the spinolaminar line (fig 2).

Principles of management include: (a) bacteriological cure; (b) correction of the osseous deformity; and (c) prevention of neurological damage.⁷ We managed our patient according to the established treatment for treating Grisel's syndrome, namely anti-inflammatory agents, muscle relaxants, physiotherapy, and a cervical collar. Patients' management should be done in consultation with a paediatric neurosurgeon. Antibiotics must be used during the acute stages of the otolaryngic infectious process. The cervical spine must be immobilised with an external orthosis given the potential risk for spinal cord injury associated with a significant degree of ligamentous laxity.^{4,7}

We have reported a symptomatic patient with a C2–3 subluxation resulting from the spread of paranasal sinus inflammation to the ligaments and joints of this cervical segment. The patient was managed with the currently used measures for treating Grisel's syndrome. We suggest that clinical and pathological manifestations in this child constitute a variant of Grisel's syndrome.

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