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Case Report

Grisel syndrome, acute otitis media, and temporo-mandibular reactive arthritis: A rare association

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ABSTRACT

We present a case report of a four-year-old boy with torcicollis and trismus after acute otitis media. Grisel Syndrome diagnosis in association with temporo-mandibular reactive arthritis was admitted, leading to early conservative treatment. GS should be suspected in a child presenting with torticollis after an upper respiratory tract infection or an ENT surgical procedure. The association with temporo-mandibular reactive findings is somehow rarer but not impossible, due to the close vascular communication between retropharyngeal and pterigoid spaces.

1. Background

By definition, Grisel's syndrome (GS) is a rare, nontraumatic subluxation of C1-C2 joint. Its name is due to the description by Pierre Grisel of three patients with pharyngitis and torticollis due to C1-C2 joint dislocation [1,2].

Although its pathogenesis remains elusive, it is plausible that those with baseline hyperlaxidity are at higher risk. That would explain why GS primarily affects children under the age of 12 years and patients with Down syndrome.

Karkos et al. [3] reported 96 cases with non-traumatic atlanto-axial rotary subluxation. Forty-eight percent occurred following infections and 40% after Ear Nose and Throat (ENT) surgery (adenotonsillectomy in78%). Within the infectious etiology, the main responsible is upper respiratory tract viral infection (83%), followed by retropharyngeal abscess (11%), otitis media (4%) and mumps (2%).

The correct diagnosis is challenging and requires a high index of clinical suspicion and appropriate radiographic imaging. Computerized tomography and/or magnetic resonance imaging of the head and neck are considered the gold standard for GS diagnosis.

The length of time until reduction has been directly related to the failure of medical treatment and to an increased risk of recurrence or permanent neck deformity [4]. Up to 15% of the untreated GS patients develop severe neurological complications: nerve damage, paralysis and even death. Early recognition is of utmost importance to avoid complications.

2. Case presentation

A previously healthy four-year-old boy was admitted to our hospital having neck pain and stiffness, with a rotational misalignment of the mental region to the right side of the neck for six days. There was no history of trauma, but the patient had rhinopharyngitis and acute otitis media diagnosed in the day prior to the beginning of the cervical complaints. He received antibiotic treatment and had remained apyretic.

The physical examination showed: torticollis, associated with spasm and tenderness over right sternocleidomastoid muscle and limited cervical range of motion (Fig. 1); trismus; right temporo-mandibular joint (TMJ) tumefaction; bilateral tympanic hyperemia; there were no asymmetry or inflammation within the tonsillar area. No neurological, ophthalmological or other osteoarticular signs were found.

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Fig. 1. Clinical presentation: right torticollis.

3. Investigation

The C-reactive protein was 2.6 mg/dL and the white blood cell count was not altered. Ultrasound examination of the neck revealed multiple enlarged lymph nodes, the biggest one measuring 22 mm × 11 mm in size and was located in the anterior jugular chain.

Contrast-enhanced CT-scan of the cervical region, showed multiple lymphadenopathies in the anterior-lateral cervical region, with no abscess detection (Fig. 2). There was a significant asymmetry of the tissues surrounding the right TMJ due to joint effusion with peripheral halo of contrast intake, suggesting of inflammatory/infectious process (Fig. 3).

4. Treatment and follow-up

First diagnostic impression lead us to assume that this was the case of a secondary torcicollis due to cervical lymphadenopaties' inflammatory process. However, initial treatment with diazepam (muscle relaxant), ibuprofen (non-steroid anti-inflammatory) and intravenous (IV) antibiotic (amoxicillin-clavulanate) lead to no further improvement.

This lead to the review of previous CT-scan images. After obtaining adequate multiplanar reconstructions for the cervical spine, additional signs of rotational dislocation of the atlantoaxial joint (Figs. 4 and 5) were identified.

After confirmation of atlantoaxial rotatory subluxation (AARS) – Type 2 dislocation according to Fielding and Hawkins classification [5], neurosurgery consulting was obtained.

In the presence of an AARS with no prior cervical trauma and only acute otitis media as the trigger factor, lead us to establish Grisel Syndrome diagnosis (Deichmueller and Welkoborsky diagnostic algorithm [6] – Table 1).

Being a type 2 AARS with less than 2 weeks of evolution, conservative management was chosen. The patient was submitted to manual subluxation closed reduction under conscious sedation by the neurosurgery team and cervical mobilization restraint was adopted using a Philadelphia collar.

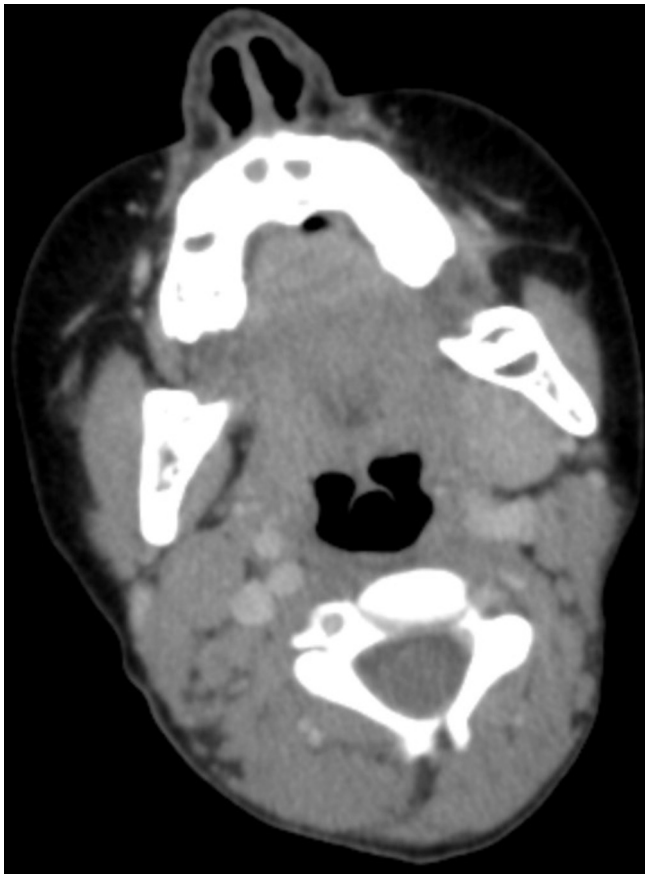

Fig. 2. Contrast-enhanced axial CT image showing multiple lymphadenopaties in the anterior-lateral cervical region, with no abscess detection.

The patient was kept on treatment with ibuprofen and IV amoxicillin-clavulanate for 11 days.

Follow-up contrast enhanced CT-scan performed after 2 weeks, and before hospital release, showed normal alignment of C1-C2, with mild edema of the atlantodental ligament (Figs. 6 and 7). No

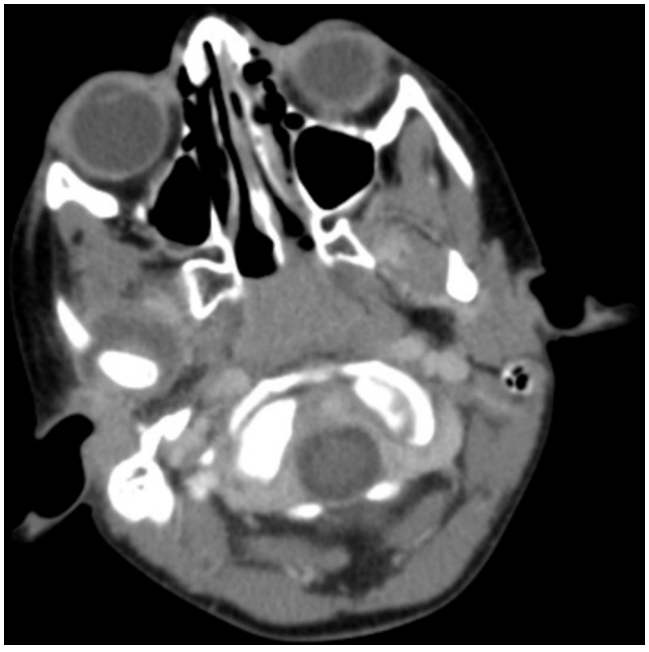

Fig. 3. Contrast-enhanced axial CT image showing right TMJ effusion.