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Case Report

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Atlantoaxial subluxation of the C1/C2 vertebrae in the pediatric patient: A case study

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Case report

A nine-year-old male presenting with neck pain and head tilt was referred for neurosurgical evaluation. According to the referring pediatrician, the head tilt and torticollis had persisted for the previous three weeks. Of note, a prodromal upper respiratory viral illness was reported, with erythema of the left mastoid area that had resolved after a few days of conservative management. The boy's family had attempted neck massages and heat compresses without resolution of the head tilt.

On examination in the office, the child was well developed and in mild distress. He had already been placed in a cervical collar by his pediatrician. The patient was unable to turn his head to the left, fixed in a "cock-robin" position (head rotated and flexed, with associated contralateral head tilt). The remainder of his physical and neurological examinations were unremarkable.

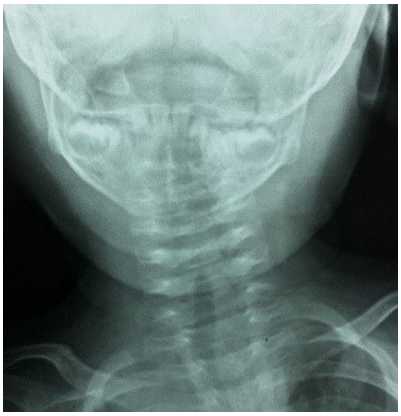


Figure 1A: Initial AP cervical spine radiograph (A) demonstrates dextroconvex rotational deformity of the cervical spine.The C1-C2 levels, however, are obscured by the patient's overlying mandible.

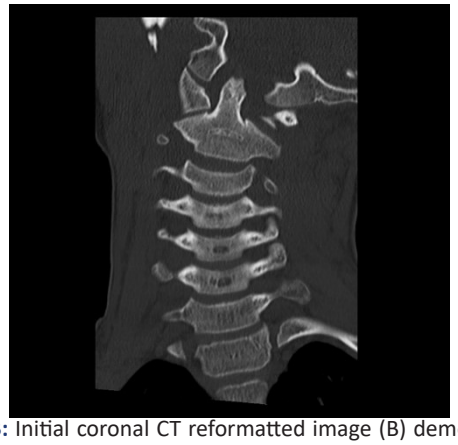
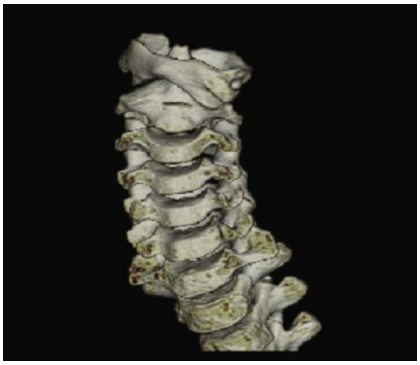


Figure 1B: Initial coronal CT reformatted image (B) demonstrates abnormal offset of the lateral masses of C1 and C2.

(C)



(D)

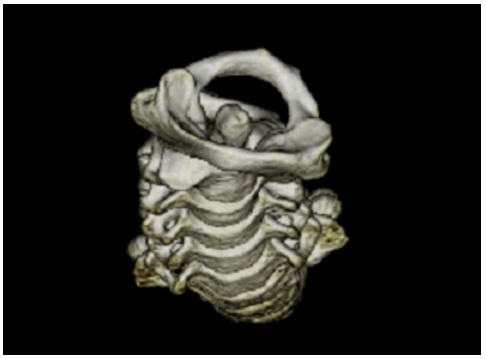


Figure 1C,D: 3D surface shaded reformat images of the CT study (C, D) best illustrate rotatory subluxation of the atlantoaxial (C1-C2) joint, with counterclockwise rotation of C1 and clockwise rotation of C2.

Follow-up MR imaging was then performed to better evaluate the C1-C2 surrounding soft tissues and ligamentous structures, and to assess for possible retropharyngeal or epidural abscess, and to exclude an underlying cervical cord mass or cord syrinx.

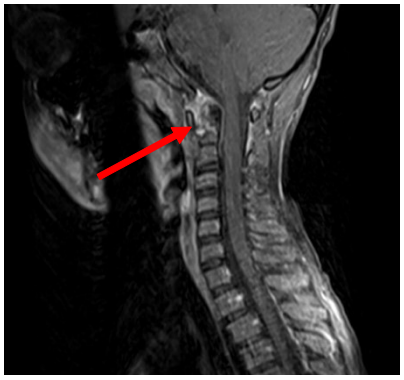
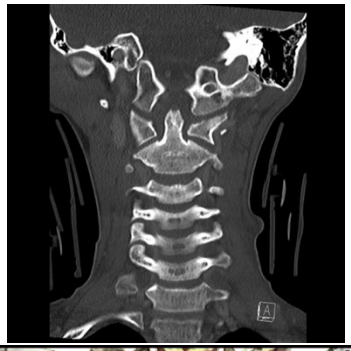


Figure 1E: Sagittal post contrast fat saturated T1 weighted sequence (E) demonstrates abnormal enhancement of the atlantoaxial ligaments including the atlantooccipital membrane, and alar and transverse ligaments due to inflammatory hyperemia (*red arrow*).

As a final attempt at manual reduction, the patient was put under an IV anesthesia with nasal airway in the operating room, with neurophysiological monitoring of the spinal cord, while the pediatric neurosurgeon performed a reduction of the cervical spine followed by the placement of a hard collar, with tape. Following the reduction, the patient was put on oral muscle relaxants.

Post-reduction imaging demonstrated the following:

(F)



(G)

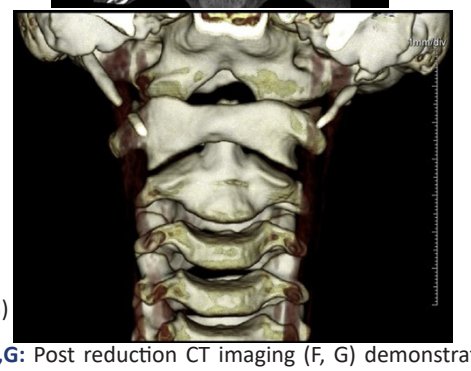


Figure 1F,G: Post reduction CT imaging (F, G) demonstrates successful reduction, with resolution of C1-C2 rotatory subluxation.

The patient was prescribed oral baclofen along with eight weeks of a hard collar followed by four weeks in a soft collar and physical therapy. With proper restrictions and care patient is expected to maintain proper cervical spine alignment and resolution of subluxation.

Discussion

Atlantoaxial rotary subluxation (AARS) is a common cervical spinal condition found in children. It involves fixed rotation or subluxation of the lateral masses of the C1 vertebrae relative to the C2 vertebrae, with patients typically presenting with painful torticollis. Proposed theories as to why it occurs more commonly in children include ligamentous laxity, more robust synovial folds, more horizontally oriented facet joints, and weak cervical musculature. Causes of AARS include trauma, postsurgical, spontaneously with underlying predisposing factors (Down syn-

drome, Morquio syndrome, Marfan syndrome), and, as in this case, inflammation after head and neck infection, referred to as Grisel's Syndrome [5].

Regional infection can cause localized irritation and neck muscle spasm, leading to non-traumatic torticollis [1]. One theory of pathogenesis purports that a pharyngovertebral to periodontoid venous network allows septic exudates to directly seed the atlantoaxial ligaments [3].

Diagnosis of AARS within three weeks of symptoms can be corrected using a cervical collar or traction, however, later diagnosis after an extended period with symptoms may require surgical correction [5].

On the cervical spine AP radiograph, AARS is commonly indicated by an asymmetric distance between the lateral mass of C1 and the C2 odontoid process. CT, however, remains the gold standard imaging modality to diagnose AARS, and can clearly demonstrate the rotatory subluxation. Follow up MR imaging can also be additive, providing soft tissue detail and possible revealing underlying infection or inflammation, and allowing for evaluation of ligamentous integrity as well as potential cervical cord compression. The presence of peridental pannus, however, has been found in greater quantities in patients with abnormal ADI. The proliferation of this tissue is most likely trigged by diseases of chronic inflammatory or degenerative nature which in turn destroy cartilage and subchondral bone. This eventually leads to instability in the cervical spine [4].

Conflicts of interest: The authors have no conflicts of interest to disclose

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