Atlanto-axial rotatory subluxation

see Atlanto-axial subluxation

Key concepts

● typically seen in children

● associations: trauma, RA, respiratory tract infections in peds (Grisel syndrome)

● often present with cock-robin head position (tilt, rotation, sl. flexion)

● Tx: early traction often successful. Treat infection in Grisel syndrome. Subluxation unreducible in traction may need transoral release then posterior fusion.

Rotational deformity at the atlanto-axial junction is usually of short duration and easily corrected. Rarely, the atlantoaxial joint locks in rotation (AKA atlantoaxial rotatory fixation [1)]

Usually seen in children. May occur spontaneously (with rheumatoid arthritis [2)] or with congenital dens anomalies), following major or minor trauma (including neck manipulation or even with neck rotation while yawning [3)], or with an infection of the head or neck including upper respiratory tract (known as Grisel syndrome [4)]: inflammation may cause mechanical and chemical injury to the facet capsules and/or Transverse Ligament of the Atlas.

Epidemiology

Atlantoaxial rotatory fixation (AARF) is a rare condition that occurs most commonly in children.

Etiology

Could occur due to congenital bony malformation, minor trauma, upper respiratory tract infections (Grisel's syndrome), postoperatively after head and neck (ENT) surgery, and unknown reasons. AARF in the postoperative patient is a rare and poorly understood entity.

A serious and often unrecognized complication of rheumatoid arthritis or ankylosing spondylitis.

The dislocation may be at the occipito-atlantal and/or the atlanto-axial articulations [5)]. The mechanism of the irreducibility is poorly understood. With an intact TAL, rotation occurs without anterior displacement. If the TAL is incompetent as a result of trauma or infection, there may also be anterior displacement with more potential for neurologic injury. Posterior displacement occurs only rarely [6)]

Classification

Fielding & Hawkins

type I: the atlas is rotated on the odontoid with no anterior displacement

type II: the atlas is rotated on one lateral articular process with 3 to 5 mm of anterior displacement

type III: comprises a rotation of the atlas on both lateral articular processes with anterior displacement greater than 5 mm

type IV: characterised by rotation and posterior displacement of the atlas vertical subluxation

Clinical features

The neurologic deficit is rare. Findings may include: neck pain, headache, torticollis—characteristic “cock robin” head position with ≈ 20° lateral tilt to one side, 20° rotation to the other, and slight (≈ 10°) flexion, reduced range of motion, and facial flattening.

The torticollis caused usually presents as abnormal posturing of the head and neck, with rotation of the chin to the opposite side.

Although the patient cannot reduce the dislocation, they can increase it with head rotation towards the subluxated joint with potential injury to the high cervical cord.

Brainstem and cerebellar infarction and even death may occur with the compromise of circulation through the VAs [7].

Pediatric emergency physicians must have a high clinical suspicion for atlantoaxial rotatory subluxation (AARS), particularly when a child presents with neck pain and an abnormal head posture without the ability to return to a neutral position. As shown in the neurosurgical literature, timely diagnosis and swift initiation of treatment have a greater chance of treatment success for the patient. However, timely treatment is complicated because torticollis can result from a variety of maladies, including: congenital abnormalities involving the C1-C2 joint or the surrounding supporting muscles and ligaments, central nervous system abnormalities, obstetric palsies from brachial plexus injury, clavicle fractures, head and neck surgery, and infection. The treating pediatrician must discern the etiology of the underlying problem to determine both timing and treatment paradigms, which vary widely between these illnesses.

Kinon et al., present a comprehensive review of AARS that is intended for pediatric emergency physicians. Management of AARS can vary widely bases on factors, such as duration of symptoms, as well as the patient's history. The goal of this review is to streamline the management paradigms and provide an inclusive review for pediatric emergency first responders [8].

Diagnosis

Radiographic evaluation

X-rays : Findings (may be confusing) include:

● pathognomonic finding on AP C-spine X-ray in severe cases: frontal projection of C2 with simultaneous oblique projection of C1. In less severe cases, the C1 lateral mass that is forward appears larger and closer to the midline than the other

● asymmetry of the atlantoaxial joint that is not correctable with head rotation, which may be demonstrated by persistence of asymmetry on open mouth odontoid views with the head in neutral position and then rotated 10–15° to each side

● the spinous process of the axis is tilted in one direction and rotated to the other (may occur in torticollis of any etiology)

CT scan: Demonstrates rotation of the atlas.

MRI: May assess the competence of the transverse ligament.

Outcome

Deformity of the superior facet of the axis (C2) and >20° of lateral inclination of the atlas observed on 3D CT are known factors in the progression to intractable [9].

Complications

The vertebral artery (VA) may be compromised in excessive rotation, especially if it is combined with anterior displacement.

Atlanto-occipital rotatory subluxation and facet deformity in the atlanto-occipital joint may occur after prolonged Atlantoaxial rotatory fixation . It is necessary to pay attention to pathological changes not only in the atlantoaxial joint but also in the atlanto-occipital joint, when treating patients with Atlantoaxial rotatory fixation [10].

Case series

In seventeen cases of irreducible atlanto-axial rotatory subluxation (here called fixation), the striking features were the delay in diagnosis and the persistent clinical and roentgenographic deformities. All patients had torticollis and restricted, often painful neck motion, and seven young patients with long-standing deformity had flattening on one side of the face. The diagnosis was suggested by the plain roentgenograms and tomograms and confirmed by persistence of the deformity as demonstrated by cineroentgenography. Treatment included skull traction, followed by atlanto-axial arthrodesis if necessary. Of the thirteen patients treated by atlanto-axial arthrodesis, eleven had good results, one had a fair result, and one had not been followed for long enough to determine the result. Of the remaining four patients, one treated conservatively had not been followed for long enough to

evaluate the result, two declined surgery, and one died while in traction as the result of cord transection produced by further rotation of the atlas on the axis despite the traction [11].

Atlantoaxial rotational fixation (AARF) is a rare entity in adults, with only a few cases reported in the English literature and often associated with a traumatic mechanism. It is an underdiagnosed condition that must be taken into account in the initial assessment of all craniocervical trauma. Both diagnostic and therapeutic delay may be a potential cause of severe neurological damage or even death of the patient. The therapeutic management is controversial given the difficulty of achieving optimum stability and permanent reduction.

The atlantoaxial rotatory fixation (AARF) and the atlantoaxial rotatory subluxation (AARS) are the most frequent manifestations of atlantoaxial rotatory dislocation (AARD) in children, and conservative treatment has proved to be suitable in many cases, considering the pathological features of these type of injuries. In literature, there is no agreement on the treatment modalities and the timing of conservative treatment.

A 28-year-old woman was involved in a traffic accident a week before coming to the emergency with rotation and irreducible cervical flexion from trauma and severe neck pain. CT and MRI column were performed and showed a cervical spinal AARF with transverse and alar ligaments intact and preserved atlantoaxial distance (Fielding I). The patient was treated by progressive cervical traction with 5 kg and manual reduction was completed in 24 h. Subsequently, an external immobilization was performed by cervical rigid collar for 16 weeks. The clinical course was good, with the patient regaining full mobility with cervical neck pain improvement.

The purpose of this paper is to show a case of a young woman with a posttraumatic AARF successfully treated conservatively. This case delineates the difficulties in diagnosing this pathology, as well as the challenges encountered in its management [12].

Case reports

A 4-year-old boy presented with atlantoaxial rotatory subluxation after a posterior fossa craniotomy to treat a cerebellar astrocytoma. Atlantoaxial rotatory subluxation was diagnosed using plain radiography and computed tomography imaging. The patient was treated with continuous cranial traction for 14 days. Initially, they detected that the patient had no C1 posterior arch or C2 spinous process; therefore, the best option was to perform the Harms technique. Postoperatively, the patient was placed in a cervical collar for 4 weeks. At the 4-year postoperative follow-up, the patient was doing well and had not developed any complications.

de Meldau Benites reported a case in which AARF can be developed after a neurosurgical procedure. Surgical techniques used for atlantoaxial rotatory subluxation should be carefully selected. The Harms technique after cranial traction was an excellent option for correcting and stabilizing the abnormal neck position. However, further studies are required to determine the best technique to use in the pediatric population [13].

A 2-year-old boy with Crouzon Syndrome undergoing posterior calvarial vault expansion (PVE) surgery developed AARF as a complication.

Results: The authors believe that cranial vault surgery should be considered a potential risk procedure for AARF, especially if it is done in susceptible populations (syndromic craniosynostosis patients) with other underlying sequelae (tonsillar ectopia or syringomyelia). During surgery, careful attention should be paid to maintaining a neutral alignment of the patient's cervical spine as rotatory movements under anesthesia and muscle relaxation may be contributory factors.

AARF should be suspected and investigated in children with painful torticollis after craniofacial surgery [14].

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