Atlantoaxial rotatory subluxation

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

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.

Types

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

Patients are usually young. 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.

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.

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 2).
