Post-Traumatic Atlantoaxial Rotatory Fixation in an Adult Presenting as Torticollis

AARF has seldom been reported in adults, and proper diagnosis is often delayed.

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tlantoaxial rotatory fixation (AARF) is a fixed subluxation or dislocation in the cervical spine involving the inferior atlantal and superior axial facets.\(^1\) This rare condition is found more frequently in children with minor trauma or upper respiratory infections,\(^2\) inflammatory conditions such as rheumatoid arthritis and ankylosing spondylitis, or predisposition to ligamentous laxity.\(^3\) AARF has seldom been reported in adults, and proper diagnosis is often delayed. Torticollis is a typical clinical feature and neurological injury is uncommon. In this case presentation, the authors present an instance of post-traumatic AARF in an adult presenting as torticollis that was properly diagnosed and treated approximately six months post injury.

**CASE PRESENTATION**

A 21-year-old female was taken to an emergency room after a severe motor vehicle accident as a restrained front-seat passenger. She was admitted to the intensive care unit for multiple injuries that included a right pubic ramus fracture, a coccygeal fracture, rib fractures, liver laceration, and a concussion. A cervical spine computed tomography (CT) scan on admission did not reveal any evidence of acute cervical spine injury.

During her 16-day hospital course, she complained of neck pain with limited range of motion. A cervical spine CT was repeated that showed no gross evidence of fracture or subluxation but a mild scoliotic curvature limiting evaluation. Further investigation by magnetic resonance imaging (MRI) revealed moderate curvature of the cervical spine with convexity to the left that was consistent with muscle spasm and no evidence of disc herniation fracture or spinal cord injury. She was diagnosed with whiplash injury and placed in a hard collar for three days followed by a soft collar on an as needed basis. She was treated conservatively for her multiple injuries and discharged to an acute rehabilitation facility for a brief period of time. As an outpatient, her symptoms persisted and radiographs at that time demonstrated cervical scoliosis. She was diagnosed with spasmodic torticollis which led to multiple ineffective treatments including prescriptions for muscle relaxants.

Figure 1: The frontal radiographs demonstrate the gross lateral displacement of the right C1 lateral mass (R C1 LM) in relation to the right superior articular facet of C2 (R C2 SAF). The lateral margins of the C1 and superior articular facets of C2 are not on the same vertical plane and there is widening of the right atlanto-dental interval.
relaxants, chiropractic care, physical therapy, and botulinum toxin injections. She was also referred to a neurosurgeon who carefully reviewed her prior cervical x-rays and recommended a cervical CT scan in a neutral, left and right rotation for suspicion of C1-C2 instability. The CT scan was completed, but she did not follow up with the neurosurgeon after her initial visit.

Approximately six months after her injury, the patient was seen at our orthopaedic center for evaluation. Our clinical findings included a left rotational fixed torticollis, significant decrease in range of motion of the neck, and normal neurological exam. Cervical x-rays taken in the office demonstrated gross lateral displacement of the right C1 lateral mass in relation to the right superior articular facet of C2 (Figure 1). The cervical CT scan (Figure 2) that was previously ordered was reviewed revealing a right C1-C2 facet joint subluxation. An MRI was ordered to further evaluate and showed a possible C1-C2 subluxation but did not identify any ligamentous injury, disc injury, or neural involvement. She was diagnosed with AARF and underwent an open reduction and internal fixation of the C1-C2 vertebrae. She was followed up three weeks later after her surgery with resolution of her torticollis and physical therapy program focusing on gentle range of motion, flexibility, and strengthening was implemented. Five months after her surgery, she remains symptom free of her torticollis.

**DISCUSSION**

AARF was first termed by Wortzman and Dewar in 1968 for its persistent asymmetrical relationship of the dens to the articular masses of the atlas unchanged by cervical rotation. This pathologic condition is rare with reports of delay in diagnosis up to twenty-eight months. This phenomenon is more frequently reported in children than adults. AARF is more commonly caused by infection or trauma, in association with a predisposing medical condition such as inflammatory or ligament laxity, or head and neck surgery. The pathophysiology is not well defined, but there appears to be a predisposition in children. The higher incidence within this population may be attributed to certain anatomical features, including a large head and underdeveloped neck musculature, rotational angle greater than 45 degrees, horizontal configuration of the atlantoaxial facets, and increased joint capsule elasticity. Even though the pathological mechanism of AARF has not been established, there have been several proposed theories including synovial inflammation producing stretching of ligaments, transverse and alar ligament ruptures, muscle spasm, ligament and capsular contractures, and articular damage. Trauma in adults such as a sports-related injury, mechanical fall, or ejection injury is usually reported as a preceding event.6-8

The atlantoaxial articulation is unique. The atlantoaxial joint is the most mobile part of the spine with the least amount of stability.6 The distinct anatomical relationship enables the majority of the left and right rotation of the cervical spine.9 The horizontal orientation of the atlas and axis facets as well as the dens pivoting on the atlas facilitates this motion.9 Stabilization of this joint is primarily by the transverse and alar ligaments which attach the dens of the axis to the posterior surface of the anterior arch of the atlas. The transverse ligament is a tight band connecting the atlas to the dens preventing anterior shift of the atlas on the axis.10 The paired alar ligaments also prevent anterior shift as well as excessive rotation.2 Other stabilizers such as the rudimentary apical ligament, the atlantoaxial ligament, anterior and posterior atlanto-occipital membranes, and atlantoaxial membrane provide minor stabilizing functions but are biomechanically insignificant.9

A common presentation in an AARF patient is painful persistent torticollis. The lateral flexion and opposite side rotation of the head or cock-robin position has been used to describe the deformity for its resemblance of a robin listening for a worm.2 Palpation of the axis spinous process may be prominent and deviated toward the direction to which the chin is pointing.7 Range of motion is significantly decreased and return to the neutral position after a rotational movement is not achieved.11 Neurological injury is uncommon but can occur, ranging from mild nerve root irritation to fatal cord compression.2,7

Unexplained persistent torticollis should be investigated. Torticollis related to muscle spasm is commonly seen, but other causes such as cervical spine infection, congenital anomalies of the dens, syringomyelia, cerebellar tumors, bulbar palsies, and ocular problems can result in torticollis. An important finding in differentiating spasmotic torticollis from AARF is that the shortened sternocleidomastoid is the force causing the deformity, while in AARF the lengthened sternocleidomastoid may be in spasm from trying to correct the deformity.2

Confirming the diagnosis of AARF is made through radiological imaging. Radiographs are typically obtained but can often be difficult to interpret resulting in an increased delay in the proper diagnosis.4,11 Lateral radiograph findings may...
demonstrate abnormal widening of space between the dens and the atlas implying transverse ligament deficiency. On open mouth radiographs, the lateral mass of the atlas that is rotated anteriorly will appear wider and closer to the midline in comparison to the opposite lateral mass.2,12

Anteroposterior radiographs may show asymmetric positioning of lateral masses of the atlas in the coronal plane.3 Computed tomography (CT) scanning is another important imaging tool to identify AARF. It is extremely useful in visualizing dislocations and fractures.13 Dynamic CT scanning can provide additional information in axial rotation of the atlas and the anatomical relationship within the occiput, atlas, and axis.14 Three-dimensional CT (3-D CT) reconstruction imaging can be beneficial, giving a global view of a cervical deformity.15 Magnetic resonance imaging (MRI) should be considered to evaluate soft tissue injury, neural involvement, disc pathology, and the presence of transverse and alar ligament tears.1,11,12 In our patient, CT and MRI were performed during her hospital course but atlantoaxial joint abnormality was not reported. This may possibly due to the resulting cervical deformity of AARF limiting full evaluation.

Treatment options include conservative care, immobilization, traction, manual reduction, and surgery.7 Conservative measures such as analgesics, collar bracing, cervical traction, or manual reduction can successfully correct the fixation.2,11 Immobilization with or without traction was advised in many cases of less than one month duration with patients achieving good long term stability.1,12 However, delay in proper diagnosis or implementation of non-surgical treatments can drastically affect its success rate. Surgery is indicated when the transverse ligament is torn or avulsed or in the presence of spinal instability, neurological injury, or failure of conservative measures.2,7 In our case, the delay in diagnosis of approximately six months with persistent neck pain, limitation of range of motion, and failure of conservative treatments prompted surgical intervention.

CONCLUSION
AARF in an adult is a rare occurrence usually related to trauma. Torticollis is a common presenting symptom, and delay of proper diagnosis can affect treatment options. AARF should be considered as a potential diagnosis in an adult with persistent torticollis after a post-traumatic event. ■

The authors have nothing to disclose.

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