Atlantoaxial Rotary Subluxation After Minor Trauma

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Abstract: We present a case of atlantoaxial rotary subluxation after a minor injury managed by manual reduction under sedation. Atlantoaxial rotary subluxation should be considered in a child with inability or unwillingness to turn their head when history and physical examination do not suggest torticollis of benign etiology. In our discussion, we review etiology, diagnosis, as well as pre- and in-hospital management.

Key Words: atlantoaxial joint, rotary subluxation, dislocation

CASE

A 5-year-old previously healthy male child was transported by emergency medical services to the emergency department with the acute complaint of severe neck pain and inability to turn his head. Upon initial assessment, he was a calm, stable, and neurologically intact child whose head was rotated 90 degrees to the left such that his chin rested nearly at his acromioclavicular joint (Fig. 1). Upon further questioning. his parents revealed that he had been wrestling with a sibling the prior evening and had struck his head against his bed frame behind his right ear. He then went to sleep and did not mention the incident to his parents until the following morning when they discovered him with his head rotated to the left with an inability to straighten it. Emergency medical services was notified. and he was transported to the emergency department on a back board with his head secured in a position of comfort (extreme leftward rotation) without a hard cervical collar in place. His medical history was unremarkable. A review of systems was negative. and there was no recent history of fever, pharyngitis, odynophagia, or upper respiratory tract infection. He was taking no medications and had no known allergies. His family history was negative for significant illnesses or neurologic disorders.

Physical examination, aside from the position of his head, was documented to be normal with full neurosensory and motor function of all 4 extremities. His temperature was 37.1°C, heart rate 128 beats per minute, respiratory rate 22 breaths per minute and unlabored, and blood pressure 103/64 mm Hg. He was calm and cooperative. His cranial nerves were grossly intact, and he had no deficits of vision, hearing, or speech. His head was normocephalic with no evidence of ecchymosis or other trauma. There was no hemotympanum. He was well perfused with strong peripheral pulses in all 4 extremities. He had no audible carotid bruits. The remainder of his secondary survey failed to demonstrate any significant injuries. He was unable to actively rotate his head to the right and passive rotation was limited by pain. He complained of significant discomfort with palpation of the left side of his neck as well as left-sided sternocleidomastoid muscle tenderness.

After a dose of oral ibuprofen, cervical spine radiographs were performed. Because of the patient's head and neck positioning, cervical spine alignment was difficult to assess on plain film and fractures were unable to be fully excluded. The films did suggest a C1–C2 rotary subluxation with abnormal leftward rotation of C1 relative to C2 (Fig. 2). A neck computed tomography (CT) scan was subsequently performed with the patient's head in the presenting orientation. Repositioning of the patient's head and neck was not attempted because of his inability to move without significant discomfort. The axial images demonstrated a C1–C2 rotary dislocation with complete loss of the normal articular contact between the C1 and C2 lateral masses (which include the transverse processes and articular surfaces) as evidenced by a 45.6-degree rotational difference between the position of anterior arches of C1 and C2 (Fig. 3). Three-dimensional reconstructed images revealed that the right C1 lateral mass was anteriorly dislocated and perched over the lateral mass of the C2 vertebrae (Fig. 4). Fractures of the cervical vertebrae were excluded. The atlas and occiput were normally articulated. No epidural or paraspinal hematomas were noted.

The patient remained stable and neurologically intact. Neurosurgery was consulted and recommended manual reduction under procedural sedation. He was transported to a room adjacent to the CT scanner, and under procedural sedation with intravenous propofol, his dislocation was reduced with gentle manual traction followed by rotation to the midline. The patient tolerated the procedure well, and a follow-up CT scan demonstrated normalization of the C1–C2 articulation. There was no further fixed rotary subluxation or asymmetry of the atlantodental interspace. There remained no fractures of the imaged vertebrae with a normal appearance of the paraspinal soft tissues as well. He was placed in an Aspen collar and allowed to awake from sedation. His postreduction neurological examination was normal. He was admitted to the hospital overnight for serial examinations and observation and was discharged the following day in good condition after requiring no further analgesia. He was instructed to remain in the cervical spine collar until reassessment by a neurosurgeon.

Two weeks later, the patient was seen in the outpatient neurosurgery clinic, and the cervical spine collar was removed. Follow-up plain radiographs were normal, and he had full range of motion of his neck. Approximately 1 month after his initial presentation, he still complained of mild residual discomfort. Therefore, a magnetic resonance imaging of the neck was ordered by his primary care physician in order to better assess his ligamentous structures. It revealed a very thin rim of signal enhancement in the interspinous tissues between the posterior elements of C1 and C2 that may have indicated a subtle ligamentous injury. Otherwise, the cervical spine was normal. He was referred to outpatient physical therapy, and has had no further complications.

DISCUSSION

Atlantoaxial rotary abnormalities have been described in the literature for over 100 years[1] with the development of a

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FIGURE 1. Photograph of our patient with head rotated ninety degrees to the left.

classification system in the 1970s.[2] Fielding and Hawkins[2] described 4 types of rotary fixation based on the degree of displacement of the odontoid process (dens) relative to the anterior facet of the atlas. The most common type, Type I, occurs with no or minimal (less than 3 mm) displacement of the dens in relation to the articular facet of the dens on the atlas and with rotation of the atlantoaxial joint within the normal range. Type II is characterized by deficiency of the transverse ligament and between 3 and 5 mm of anterior displacement of the atlas. The amount of rotation is in excess of that normally achieved by the C1–C2 articulation. Type III is defined by anterior displacement of the atlas by more than 5 mm with associated weakness of the transverse and secondary ligamentous structures. Type IV is rare and has been described mainly in adult patients with rheumatoid arthritis. It was related to posterior displacement of the lateral masses of C1 in relation to the dens.

The primary function of the atlantoaxial joint is rotation rather than flexion or extension.[3] The joint is stabilized by the transverse ligament (which limits excess flexion) and the alar ligaments (which limit excess rotation).[3] The articulation between the first and second cervical vertebrae in children displays unique anatomical features such as an inherent ligamentous laxity that may lead to impingement of the meniscus-like synovial folds at the lateral atlantoaxial joints.[4,5] This may explain the predominance of atlantoaxial rotary subluxation-fixation cases in children. There have been relatively few studies that have clearly defined the epidemiology of atlantoaxial rotary abnormalities. Though uncommon, most cases in the literature, especially those attributed to injury, have occurred in children.[3,5–7] Minor trauma is a known antecedent risk factor, but several other causes have also led to documented cases of atlantoaxial rotary subluxation/dislocation. Grisel syndrome is an oft described clinical entity with a nebulous pathologic explanation.[8,9] It is theorized to occur as a result of ligamentous laxity of the C1–C2 joint secondary to an inflammatory or infectious process in the head and neck.[8] Cases have been described in patients with otitis media, retropharyngeal abscesses, lupus, rheumatoid arthritis, and in patients that have undergone otolaryngologic procedures.[10]

Evaluation for involvement of the spinal cord is the first priority in any patient with neck stiffness or malposition. The cervical spine should be properly immobilized at all times. If spinal cord signs and symptoms are present, emergent imaging and neurosurgical consult are warranted.

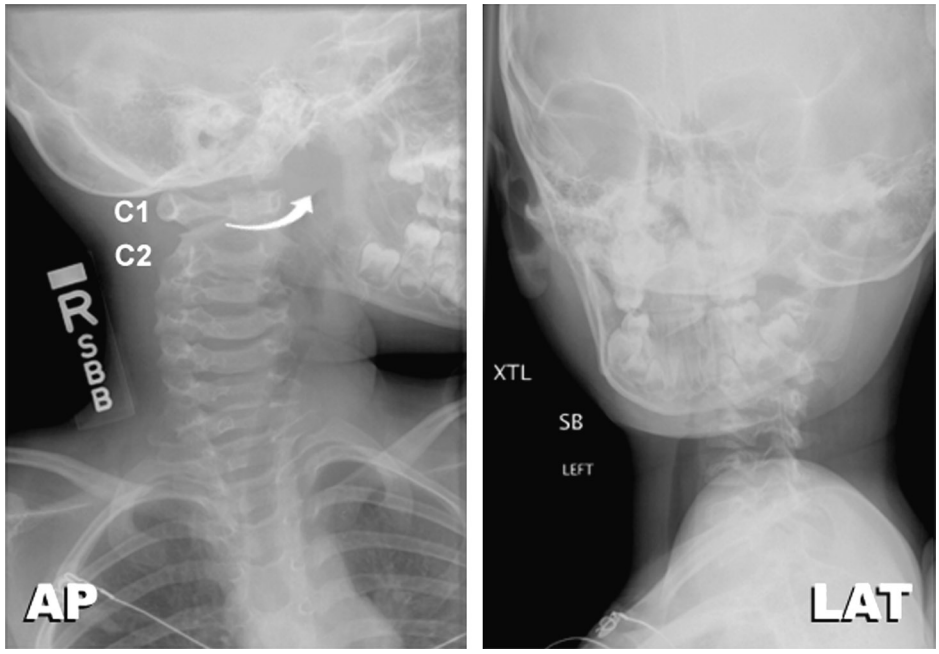

FIGURE 2. Anterior-posterior and lateral radiographs suggesting C1–C2 rotary subluxation.

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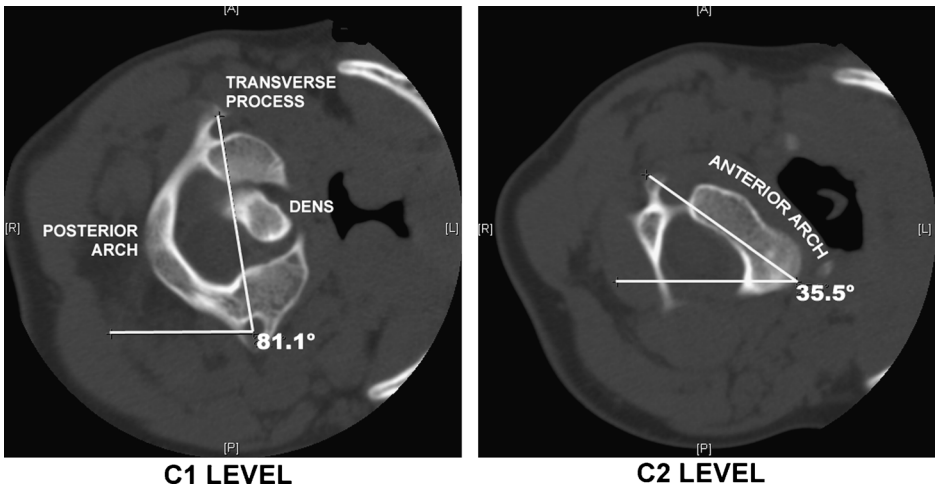

FIGURE 3. Axial CT with angle of rotation at the level of C1 and C2.

Clinicians should then assess for a history of trauma including mechanism of injury. If the patient is febrile, assessment for meningismus, cervical lymphadenopathy, or other signs of infection in the head and neck is paramount.

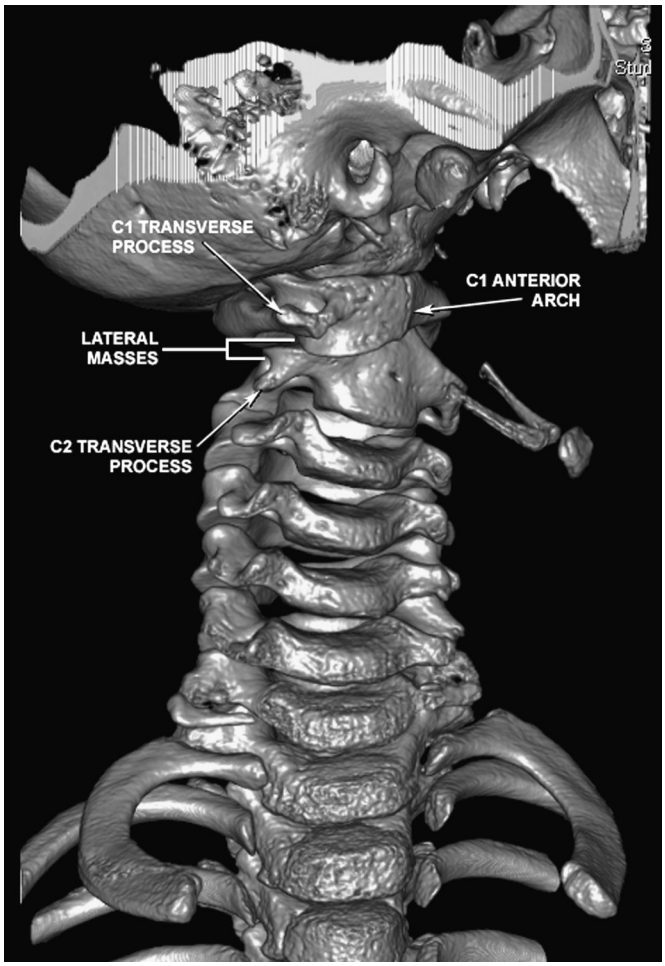

FIGURE 4. Three-dimensional reconstructed CT image showing atlantoaxial subluxation.

A headache could indicate an intracranial process such as a mass or subarachnoid hemorrhage and may warrant imaging. Location of a neck mass in an afebrile child with torticollis will help to differentiate between congenital muscular torticollis (if in the sternocleidomastoid of an infant), or more serious causes such as malignancy or atypical infection. It is also important to consider medical history, especially for inflammatory disorders, preexisting conditions (Down syndrome, Klippel-Feil syndrome, Chiari malformation), as well as recent surgery of the head and neck. In general, simple cervical muscle strain is a diagnosis of exclusion.[11]

The diagnosis of C1–C2 rotary subluxation should be considered when a patient's symptoms and presentation do not fit those of torticollis because of a muscular or ligamentous strain. The head is often held in the classically described "cock robin" position with the head tilted (laterally flexed) to 1 side and the head rotated to the side opposite the facet dislocation.[2] The head is often rotated to an extreme degree.[3] The spinous process of C2 can be rotated in the same direction as the head and is sometimes palpable.[12] In the absence of severe trauma, most patients will have a normal neurologic examination.[11] Severe pain and an inability and/or unwillingness to move the head will regularly be present. Though spasm of the sternocleidomastoid muscle is present in many causes of torticollis, midline muscle stiffness is a more worrisome sign.[11] Important attention must be paid to the acuity of onset as well as the duration of illness. Most cases of torticollis due to a muscular or ligamentous etiology resolve in 5 to 7 days and are easily managed with analgesic and antiinflammatory medications, a soft cervical collar for comfort, and close follow-up.[11,13]

Diagnosis is achieved through a careful history and physical examination with the additional aid of radiologic evaluation. Plain radiographs of the cervical spine are often the first study performed but are not always revelatory because of head positioning and limitation of range of motion. Head tilt often makes the lateral film difficult to interpret because it obscures the normal anatomical position and

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landmarks of the cervical vertebrae. Furthermore, an odontoid (open mouth view) is best obtained in the anterior-posterior position which may not be achievable in these patients. If the child presents with extreme rotation or pain, it is reasonable to attempt films in the position of comfort only in order to avoid further injury. The availability of other imaging modalities can obviate the need to obtain necessary diagnostic information from plain radiographs alone. Interestingly, atlantoaxial rotary abnormalities may also present in a child without an obvious abnormality of head rotation.[13] Therefore, CT is the imaging modality of choice. It is readily available and affords essential information in the assessment of abnormalities of the cervical spine.[13,14] Axial CT images can allow the radiologist to quickly assess for abnormalities of the position of the dens relative to the anterior facet of the atlas. Computed tomography may also reveal fractures not seen on plain radiography. Newer techniques such as 3-dimensional reconstructions of the cervical spine provide dramatic images that greatly aid in diagnosis.[7,13,15] Magnetic resonance imaging eliminates the risk of ionizing radiation but is more costly and difficult to obtain. It does, however, afford better resolution of the ligamentous structures and spinal cord when compared with CT.[13]

Prehospital care is an important consideration in patients with atlantoaxial rotary subluxation. Emergency medical services personnel are taught through the pediatric education for prehospital professional course as well as through traditional prehospital education texts that certain contraindications exist for moving a patient's head to an in-line position before transport. The pediatric education for prehospital professionals course warns against forcing neutral position if there is resistance to movement, crepitus, or increased spinal pain.[16] Mosby's Paramedic Textbook lists resistance to movement, neck muscle spasm, increased pain, the presence or increase in neurological deficits during movement, and severe misalignment of the head away from the midline of the body axis as contraindications to manual in-line immobilization.[17] In these cases, the patient should be transported with the head secured in a position of comfort so as to avoid incurring new or additional neurologic injury.

Management of atlantoaxial rotary subluxation ranges from conservative measures to invasive surgical procedures. Manual reduction may be indicated in patients with recent onset of symptoms.[6,18] In a retrospective study with a small cohort of patients, Subach et al[12] found that most cases resolved quickly with closed reduction followed by mechanical traction, either via a hard cervical collar or weighted traction. Some did experience a recurrence of the subluxation and ultimately required surgical correction via posterior fixation. Although there are no specific recommendations, a soft cervical collar is often worn after reduction until patients are pain-free and have normal follow-up cervical radiography.[18] Our patient was manually reduced under procedural sedation with propofol and then placed in an Aspen collar without traction. There are no studies, however, that compare outcomes of manual reduction versus traction. Overall, most children have an excellent prognosis after reduction although some do ultimately require surgery.[12,18] Most of the studies that have followed children long term are concerned with posterior fusion and its outcomes rather than nonoperative management. In patients that are fused, bone degeneration is a concern in as little as 10 years.[19] Another concern is limitation of range of motion after surgical fixation. There is little information about prognostic factors that are used to guide potential therapy. The length of time that the subluxation has been present may directly correlate with worse outcome and increased relapse rate.[5,12,18] In a retrospective review of 20 children with reduced rotary atlantoaxial subluxation collected over a period of 7 years, Subach et al[12] noted that conservative management was successful in 14 cases. However, 6 required posterior fusion. Two had not been reduced by cervical traction, and 4 had multiple recurrences. In general, those that had recurrent subluxation had a mean duration of symptoms, including pain and limitation of neck movement of 69.7 days before initial diagnosis and management compared with 13.8 days for those in which subluxation did not recur. Based on their data, they felt that a period of 3 weeks or longer of persistent subluxation was associated with the greatest risk of relapse.

Atlantoaxial rotary subluxation should always be considered when the history and physical examination do not suggest torticollis of benign etiology. A thorough physical examination is essential in order to evaluate for potential etiologies and for neurologic deficit. Diagnosis is most effectively achieved through a combination of careful history and radiography. Newer CT imaging modalities, such as 3-dimensional reconstruction, have greatly aided in the evaluation of atlantoaxial rotary abnormalities. Though controversy exists with regard to management, most patients recover with limited or no significant sequelae. The most important factor in promoting a good outcome is early diagnosis and management.

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