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Congenital laxity of the transverse atlantal ligament producing atlanto-axial subluxation: a case report

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An anterior atlanto-dental interspace (ADI) of greater than 3mm in an adult indicates atlanto-axial subluxation (AAS) and instability. A number of pathological processes are known to cause AAS. Trauma, specifically injuries with a component of hyperflexion, may produce sprain or avulsion of the transverse atlantal ligament (TAL). More often, however, the odontoid process will fracture before the tough TAL ruptures, so that isolated rupture of the ligament is generally considered to be rare. Inflammatory arthritis or infection may erode and weaken or destroy this ligament. Focal or global congenital conditions are also associated with TAL laxity or agenesis and subsequent AAS. Congenital laxity of the TAL is a diagnosis of exclusion which may be made only after a comprehensive search for other causes of AAS. (JCCA 1988; 32(4): 187–190)

KEY WORDS: abscess, atlanto-axial subluxation, atlanto-dental interspace, Down's syndrome, psoriasis, rheumatoid arthritis, spinal injuries, transverse atlanto-axial ligament, chiropractic, manipulation.

Un intervalle atlanto-dental antérieur (ADI), supérieur à 3mm chez l'adulte, est un signe d'instabilité, et révèle une sous-luxation atlanto-axiale (AAS). Plusieurs processus pathologiques ont été reconnus comme pouvant être la cause d'une AAS. Un trauma, et particulièrement des blessures comportant un facteur d'hyperflexion, peuvent entraîner un étirement ou une avulsion du ligament atlantal transverse (TAL).

Cependant, il arrive plus souvent que le processus odontoïde se fracture avant que le solide TAL ne rompe, de sorte qu'une rupture isolée du ligament est généralement considérée comme un fait rare. De l'arthrite inflammatoire ou une infection peuvent éroder et affaiblir ou même détruire ce ligament. Des conditions congénitales focales ou globales sont également associées à la laxité ou l'agénésie du TAL, et à la AAS qui en résulte. Une laxité congénitale du LAT est un diagnostic d'exclusion qui ne devrait être établi qu'après recherche complète des autres facteurs ayant pu causer la SAA. (JCCA 1988; 32(4): 187–190)

MOTS-CLÉ: abcès, sous-luxation atlanto-axiale, intervalle atlanto-dental, syndrome de Down, psoriasis, polyarthrite, blessures à le colonne vertébrale, ligament atlanto-axial transverse, chiropraxie, manipulation.

Introduction

Several references to isolated congenital laxity of the TAL have appeared in the literature.[1–6] More often, however, laxity is discussed within the context of an associated pathology. Specific figures concerning the incidence of congenital laxity or agenesis of the TAL are available only in association with Down's syndrome, in which a reported 20% of individuals are affected.[7,8] The majority of these cases remain asymptomatic and are discovered only upon radiologic survey.[7,9,10] This may suggest that congenital laxity of the TAL unassociated with other disease is more common than published reports indicate.

This paper will discuss the relevant literature and present a case report of congenital laxity of the transverse atlantal ligament.

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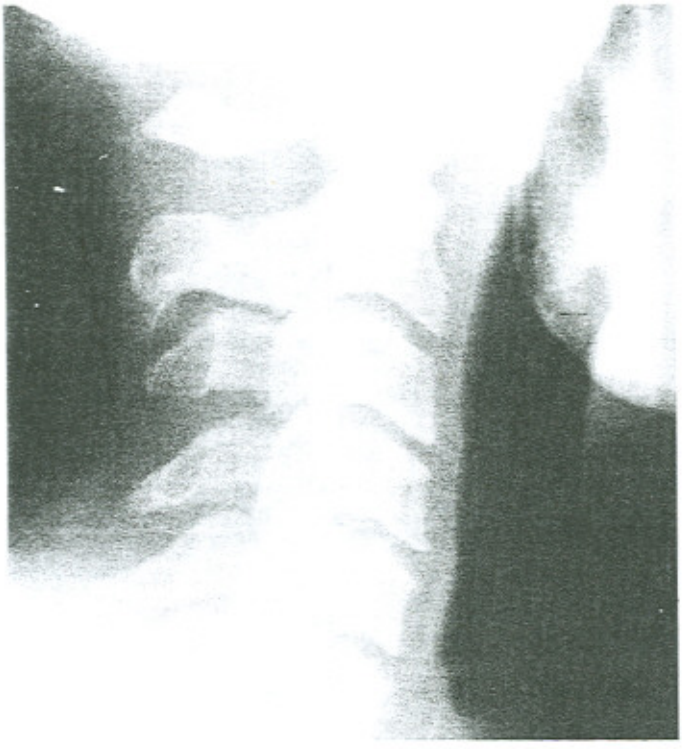
Case report

A 26-year old white male construction worker presented to a chiropractic office complaining of generalized neck pain and dizziness upon rotation of the head to the left. These symptoms had been present for approximately one year, with increasing frequency and severity over the past two months. Periods of "blackouts" and neck pain so severe that the patient was unable to turn his head in either direction were occurring twice a month, with each episode lasting about one week. The onset of these symptoms could not be related to a specific traumatic incident, but the patient did admit to repeated mild "overuse" of his neck and upper extremities during the course of his work. A systems review failed to reveal evidence of congenital anomaly, inflammatory joint disease, or infection, either local or systemic.

The physical examination revealed painfully restricted motion of the cervical spine, particularly in extension and left rotation. Tests for vertebro-basilar insufficiency were performed twice during the course of the exam because of the



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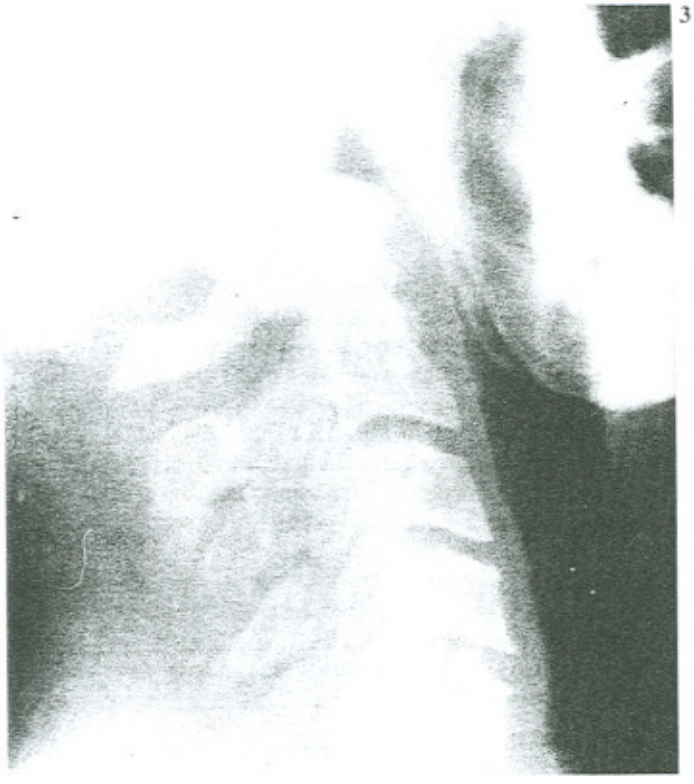

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Figure 1–3 Atlanto-axial subluxation is demonstrated by an increase of the atlanto-dental interspace between extension and flexion. The ADI increases to 4.0mm during flexion. A subtle anterior translation of the posterior cervical line (spino-laminar line) between extension and flexion provides further evidence of instability.

patient's history of dizziness during head rotation. The tests were negative. The remainder of the examination was unremarkable.

Radiographic examination of the cervical spine demonstrated an increased atlanto-dental interspace (ADI) ranging from 2.0mm at the caudal aspect of the interspace to 6.0mm at the cephalad margin. The midpoint measurement was 4.0mm. (Figure 1) Because the stability of the upper cervical complex was in question, an unforced flexion view was obtained under careful scrutiny for signs and symptoms of spinal cord compression. The flexion lateral view demonstrated no change in the ADI from that noted on the neutral film. (Figure 2) Extension of the cervical spine produced uniform reduction of the ADI to 1.0mm. (Figure 3)

The remainder of the radiographic examination was unremarkable. It is important to note that C1 existed as a complete and freely mobile segment with no evidence of assimilation to the occiput. Other pertinent negative findings included normal prevertebral soft tissues (which mitigates against traumatic or

infectious etiology) and lack of instability in the remainder of the cervical motion segments.

A computed tomography (CT) scan was performed, revealing a 2.5–3.0mm focal left-sided posterior disc protrusion at C4–5. While this may have accounted for some of the patient's neck pain, it was felt to be unrelated to the symptoms of vertigo and syncope. The CT scan was otherwise unremarkable. A magnetic resonance (MR) scan was then performed, which verified the findings on the CT scan but demonstrated no other abnormality. Because of the nature and severity of the symptoms, an electromyogram (EMG) of the upper extremities and diagnostic ultrasound (US) of the vertebral and carotid arteries were obtained, both of which were negative.

A trial of conservative therapy was begun, including high velocity, low amplitude spinal manipulation utilizing short levers, along with non-specific mobilization. Therapy was directed to the mid- and lower cervical spine, avoiding the upper cervical complex. This was combined with electrical muscle stimulation (EMS), high volt galvanic (HVG) and transcutaneous electrical nerve stimulation (TENS) for reduction of pain and spasm. When the patient's response to this treatment was unsatisfactory, he was referred to an eye, ear, nose and throat (EENT) specialist, whose examination provided no further insight.

The patient was subsequently referred back to his chiropractor, where the previous treatment regimen was resumed, with the addition of therapeutic exercises. The patient showed gradual improvement, with reduction in the intensity and duration of his neck pain and "dizzy spells" over a period of several months. A 75–80% improvement was reported by the patient at the time of discharge, a clinical course reported to have been satisfactory to both the patient and physician, in light of the severity and duration of symptoms at the time of presentation.

Table 1 Causes of Atlanto-axial subluxation

1 Traumatic
   a. Stretched or ruptured TAL
   b. Odontoid fracture
2 Congenital Anomalies of the Cervical Spine
   a. TAL laxity or agenesis
      1) Isolated
      2) Associated with Down's syndrome
      3) Associated with Neurofibromatosis (rare)
   b. Atlanto-occipital assimilation ("Occipitalization" of the atlas)
      1) Isolated
      2) Associated with Klippel-Feil syndrome
   c. Odontoid agenesis or hypoplasia
      1) Isolated
      2) Associated with a dysplastic syndrome (e.g. Morquio syndrome)
   d. Agenesis of the posterior arch of atlas
3 Inflammatory Arthritis
   a. Rheumatoid arthritis
   b. Psoriatic arthritis
   c. Juvenile Chronic Arthritis
   d. Ankylosing Spondylitis
   e. Systemic Lupus Erythematosis (SLE)
   f. Reiter's syndrome
4 Infection (Retropharyngeal Abscess)
5 Conditions of Generalized Ligamentous Laxity
   a. Marfan syndrome
   b. Ehlers-Danlos syndrome
6 Tumor (Primary or Metastatic)

Discussion

The causes of an increased ADI in an adult are numerous and varied. These are summarized in Table 1. The clinician must be thorough in ruling out each possibility. Only after this is done may a diagnosis of congenital laxity of the TAL be made.[1,2]

Pathophysiology

The motion of C1 on C2 during flexion of the cervical spine is completely dependent upon the integrity of the surrounding ligamentous and capsular structures.[2] The anterior shift of C1 on C2 will not exceed 3mm in an adult if the TAL is intact. An ADI of 5mm is indicative of ligamentous rupture.[11] It is in this range that the secondary supporting ligaments, specifically the alar and apical ligaments, begin to exert a checkrein effect, providing the "second line of defense" in preventing spinal cord compromise.[12] It is important to note that the only specific references in the literature regarding where the measurement of the ADI should be taken are in the form of diagrams; but, based upon these drawings it appears that most authors make the measurement between the midpoint of the posterior aspect of the anterior tubercle and the anterior aspect of the dens.

Steele provided insight into the relative paucity of symptoms despite the presence of AAS through his anatomic studies of the upper cervical complex. He developed a "rule of thirds" which states, simply, that the area of the spinal canal at C1 may be divided from anterior to posterior into one-third odontoid process, one-third spinal cord and one-third "space." This "space" is composed primarily of relatively compressible loose areolar and connective tissues and is approximately equal to the transverse diameter of the odontoid. This region provides a margin of safety in the presence of AAS.[12] In general, an individual will not experience symptoms of cord compression until the ADI has increased beyond this "safe zone", the size of which varies greatly from person to person.[11] This represents an individual's "first line of defense" against spinal cord compromise.[12]

The limited symptomatology seen in this case is not exceptional. A reported 20% of individuals with Down's syndrome have laxity or agenesis of the TAL[7,8], but the majority of cases remain asymptomatic and are discovered only upon radiologic survey or following trauma.[7,9,10] It appears that the most critical factor in the generation of symptoms is the space available for the spinal cord (SAC).[13] This space extends from the posterior aspect of the odontoid process of C2 to the anterior surface of the posterior arch of C1. In general, the SAC must be decreased to 13mm or less before neurologic deficit is produced.[14] The SAC in this patient was a generous 23mm.

Treatment

References to treatment considerations of isolated congenital laxity of the TAL have not been made. It may be useful to review the treatment of Down's syndrome patients with TAL laxity or agenesis. Although prophylactic fusion of the craniovertebral junction has been performed, the results have been

inconsistent; and, it is suggested that in the absence of symptoms of cord compression, fusion is contraindicated.[2] External immobilization or other conservative approaches have not been studied, but it seems prudent to suggest that manipulative therapy to the upper cervical spine is absolutely contraindicated. While a definitive statement regarding the efficacy of chiropractic management of congenital AAS may not be made based upon this single case report, the positive results obtained suggest that further research in this area may be warranted. It must always be borne in mind that in the presence of AAS significant cord compression may occur even in the absence of non-physiologic or manipulative motions.[15–18]

The "V"-shaped Predens Space (PDS)

In 1985, Bohrer, et al described a "V"-shaped appearance of the predens space (atlanto-dental interval) in patients involved in acute cervical spine trauma.[19] They felt that while this may be due to tearing of the relatively weak upper cervical posterior ligaments, leaving the transverse ligament intact, it was more likely related to a non-traumatic developmental cause. It appears that some individuals simply show a disproportionate amount of flexion of C1 on C2.

While there is a superficial similarity between the cases described by Bohrer and colleagues and the case reported here, significant differences exist, making it likely that this case is unique. The patient in this report was a symptomatic 26-year old with no recent trauma, while the 4 cases of Bohrer, et al, were isolated from among 26 emergency room patients, all of whom experienced an acute cervical spine injury. The ages of these patients ranged from 9–18 (average 14.75), a much younger population.

Bohrer and coworkers found that the caudal measurement of the predens space (PDS) ranged from 0–1.5mm and that the interspinous distance (ISD) between C1 and C2, measured from the inferior aspect of the posterior tubercle of C1 to the superior aspect of the C2 spinous process, increased 14–17mm between the extremes of extension and flexion.[19] The case reported here showed a caudal ADI of 2.0mm, yet the interspinous distance increased only 6mm between extension and flexion. This suggests true AAS from TAL laxity allowing forward translation of C1 on C2, rather than merely disproportionately greater flexion of C1 on C2. Bohrer, et al, also found that most patients had a predens space (PDS) angle of less than 11 degrees (maximum 21 degrees) and that most patients with a PDS angle greater than 11 degrees had a physiological posterior tilt of the dens. The patient in this case report had a PDS angle of 23 degrees, yet had no posterior tilt of the dens.

Table 2

	Mick	Bohrer, et al[19]
Age	26 yo	9–18 yo (ave 14.75)
Trauma	No	Yes
Symptoms	Yes	No
Caudal ADI (PDS)	2.0mm	0–1.5mm
Increase in ISD	6mm	14–17mm
PDS Angle	23 degrees	Most <11 degrees
Posterior Tilt of Dens	No	Yes

The differences between the cases reported by Bohrer, et al, and this case have been summarized in Table 2. It is felt that the differences are sufficient to indicate that this case is unique from those reported earlier.

Conclusion

A case has been presented of a young adult male with atlanto-axial subluxation, having no recognizable etiology. In the absence of a significant identifiable traumatic episode or underlying pathology, a diagnosis of congenital laxity of the transverse atlantal ligament has been advanced. Except in individuals with Down's syndrome, this entity has received limited attention in the literature. Study of a collection of similar cases might provide insight into the pathogenesis, incidence, and management of this entity.

References

1 Greenberg, AD. Atlanto-axial dislocations. Brain 1968; 91: 655–684.
2 Hensinger, RN. Osseous anomalies of the craniovertebral junction. Spine 1986; 11(4): 323–333.
3 Frank, I. Spontaneous (non-traumatic) atlanto-axial dislocation. Ann Otol, Rhinol, & Laryngol 1936; 45: 405.
4 Titrud, LA, McKinlay, CA, Camp, WE, Hewitt, HB. Non-traumatic atlanto-axial dislocation. J Neurosurg 1949; 6: 174.
5 Makon, RF, Lovell, WW. Spontaneous atlanto-axial subluxation accompanied by severe neurologic deficits. Surgery 1956; 40: 770.
6 Rothman, RH, Simeone, FA. The Spine, 2nd ed. Philadelphia: WB Saunders, 1982: 194–200.
7 Martel, W, Tishler, JM. Observations on the spine in mongoloidism. Am J Roentgenol 1966; 97: 630–638.
8 Spitzer, R, Rabinowitch, JY, Wybar, KC. A study of the abnormalities of the skull, teeth and lenses in mongoloidism. Can Med Assoc J 1961; 84: 567–572.
9 Curtis, BH, Blank, S, Fisher, RL. Atlanto-axial dislocation in Down's syndrome. JAMA 1968; 464–465.
10 Dzenitis, AJ. Spontaneous atlanto-axial dislocation in a mongoloid child with spinal cord compression: a case report. J Neurosurg 1966; 25: 45, 460.
11 Fielding, JW, Cochran, GV, Lawsing, JF III, Hohl, M. Tears of the transverse ligament of the atlas. J Bone Joint Surg 1974; 56A: 1683–1691.
12 Steele, HH. Anatomical and mechanical considerations of the atlanto-axial articulations. J Bone Joint Surg 1968: 50: 1481–1482.
13 McRae, DL. Bony abnormalities in the region of the foramen magnum: correlation of the anatomic and neurologic findings. Acta Radiol 1953; 40: 335–354.
14 Spierlings, ELH, Braakman, R. Os odontoideum: analysis of 37 cases. J Bone Joint Surg 1982; 64B: 422–428.
15 Stratford, J. Myelopathy caused by atlanto-axial dislocation. J Neurosurg 1957; 14: 97–104.
16 Wadia, NH. Myelopathy complicating congenital atlanto-axial dislocation. Brain 1967; 90: 449–472.
17 Finerman, GA, Sakai, D, Weingarten, S. Atlanto-axial dislocation with spinal cord compression in a mongoloid child. J Bone Joint Surg 1976; 58A: 408–409.
18 Nordt, JC, Stauffer, ES. Sequelae of atlanto-axial subluxation in two patients with Down's syndrome. Spine 1981; 6: 437–440.
19 Bohrer SP, Klein A, Martin W. "V"-shaped predens space. Skeletal Radiology 1985; 14: 111–116.