Odontoid agenesis with atlanto-axial luxation

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Atlanto-axial luxation was first described in 1830 and since that time over 200 cases have been reported in the world literature. All of these patients are at risk, either from minor trauma or from progressive cord myelopathy. Symptoms vary greatly and range from neck pains and stiffness to transient quadriparesis. Specific chiropractic adjustment was successfully employed in this instance, however, grave consequences could result if manipulative procedures were attempted without adequate x-ray studies.

KEY WORDS: atlas, luxation, dens agenesis, manipulation, chiropractic.

La luxation atlas-axis a été décrite pour la première fois en 1830; depuis cette date, la bibliographie mondiale en a enregistré plus de 200 cas. Tous ces patients risquent, soit des traumatismes mineurs soit une myléopathie de la moelle épinière. Les symptomes sont très variés et vont de douleurs et d'une raideur du cou à une quadriparésie transitoire. Dans ce cas, on a eu recours à un traitement spécifique par chiropraxie, mais on risque des conséquences graves si les procédures de manipulation sont enreprises sans un examen adéquat aux rayons-X.

MOTS CLÉS: atlas, luxation, agénésie, manipulation, chiropraxie.

Case report

This 29 year old male's presenting complaint was a right-sided, mid-cervical spine pain, pain over the right acromioclavicular area, as well as mid-thoracic spine pain. Neck pain and stiffness was especially troublesome first thing in the morning. Sneezing intensified the neck pain. The most recent episode started the previous day while in an awkward position in a crawl space. While reaching to install a section of heating duct work, a "clicking" sound was heard in the neck with immediate discomfort being felt.

Four years previously the patient had been in a motor vehicle accident wherein his truck rolled over. After the accident he complained of a sore neck for a few days, but otherwise was unhurt. Later, while working as a motor mechanic, he experienced an occasional sore neck if his work involved heavy lifting.

His general health was good and he was not on any prescribed medication.

Examination of this 5'11" slightly built male revealed a gentle "S" scoliosis from C1 to L5 with a posterior gravity line of 1. Range of motion of the cervical spine was limited and painful on extension (0), right side-bending ($^1/_2$), and right rotation (50°), with pain localized over C6 on the right side. The upper extremity reflexes were +1. Tenderness to deep palpation was localized over C6 on the right, T5, T7, T8, and T9 on the left side. The thoracic spine tenderness was unrelated to the presenting complaint.

Stress films of the cervical spine are quite remarkable. There is a congenital anomaly of C2 with an absence of the odontoid. Marked instability is present at the atlanto-axial motion segment. The extension film (Figure 1) indicates normal alignment of C1-C2 while the neutral film (Figure 2) and the flexion film (Figure 3) show a forward displacement of the atlanto-axial joint by 22 mm. The tomogram (Figure 4) clearly reveals the absent odontoid peg.

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Hunter's[1] paper in 1968 stated that displacement of the atlanto-axial joint was first described by Bell in 1830 and since that time over 200 cases have been reported in the world literature. He is of the opinion that the condition is more common than was initially assumed.

With the remarkable instability that this patient exhibited, he was referred to an orthopedic surgeon who recommended surgical fixation of the area as it was felt that the patient's neck was at risk. According to Sherk[2], Weigle[3], Fromm[4], and Hunter[1], most authorities now recommend surgical stabilization because of the probability of developing a progressive permanent myelopathy. It is felt by Fromm[4] and Hunter[1] that progressive and repeated dislocation of the atlas on the axis produces meningeal hypertrophy and irritation which in turn compromises the cord blood supply with resultant demyelination and possible death. Autopsies on similar cases showed marked spinal cord softening at the level of the narrowing of the spinal canal, with demyelination and intense microglial reaction seen microscopically. It appears, according to Fromm[4], that the resulting neurological deficits are apt to be irreversible once they do appear. To the best of my knowledge my patient refused the surgeon's advice.

Discussion

It is difficult to understand why this man has never had any cord signs or symptoms. Congenital absence of the odontoid process was first described by Roberts[5] in 1933. Garber[6], in a small group of patients with congenital conditions of the upper cervical spine, found that these patients exhibited symptoms of neck pain and stiffness, transitory quadriparesis, progressive hemiparesis (one patient) and headache. However, he also found that head and neck trauma precipitated the symptoms, otherwise the abnormalities would have gone unnoticed. The severity of the symptoms, when present, according to Garber[6], seemed out of proportion to the moderate gravity of the injury. Sherk[2] states that the majority of these people show symptoms in the second or third decade of life usually after minor injury. In Sherk's[2] opinion an adequate diagnostic work-up usually

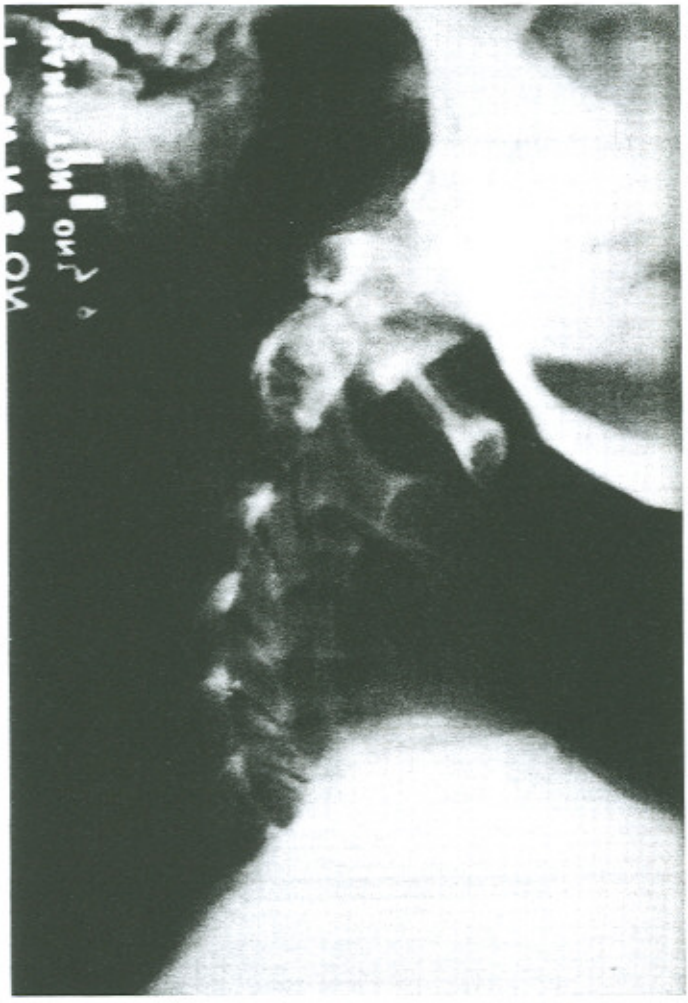

Figure 1: Extension view shows normal alignment of C1-C2.

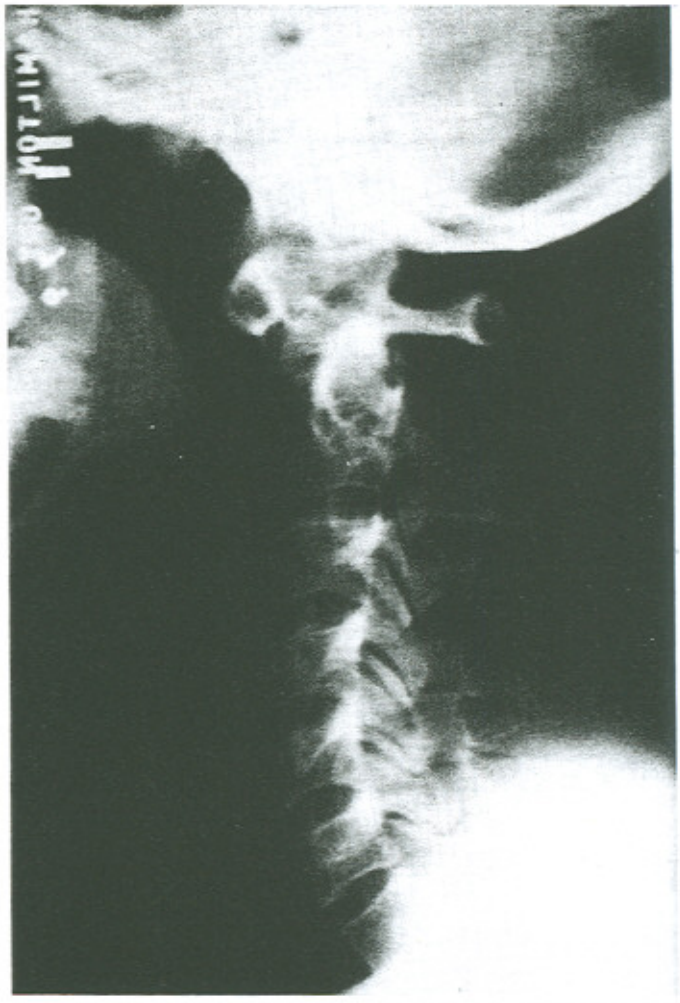

Figure 2: Neutral view shows forward displacement of atlanto-axial joint by 22 mm.

requires, in addition to the routine cervical spine views, flexion-extension films, tomography, cineradiography and contrast studies.

Fromm[4] reported that in 27 known cases, six developed progressive cervical myelopathy while five developed post traumatic myelopathy or radiculopathy. Two of his own patients presented with progressive spastic quadriparesis of obscure etiology, with symptoms of recurring urinary retention, and recurring numbness in both hands and feet, and positive Rhomberg's and Babinski signs.

Wollin[7] describes 14 cases of forward luxation of the atlas in which some patients had no history of injury or had only a minor history of injury. Infection didn't account for any of his reported cases. Six of the cases had associated congenital cervical spine abnormalities and Wollin[7] suggests that underdevelopment of the cruciate ligament could be a cause of atlas dislocation. McRae[8] reached a similar conclusion when three of his six cases had associated congenital bony anomalies. Part of Wollin's[7] study reached the conclusion that there is no correlation between the degree of reduction of the spinal cord space at the level of the ring of the atlas and the presence of neurologic signs. The space available for the spinal cord within the spinal canal varies in each individual and this is an important determining factor in the development of neurological symptoms.

Similarly, Gehweiler's[9] patient in the only known case of spondylolisthesis of the axis, experienced neck pain lasting one to three days, followed by remissions lasting three months, with only a few experiences of numbness in the left upper limb.

Althoff's[10] experiments with cadaver cervical spines found that the ligaments of the atlanto-axial joint act as a uniform body and that cutting of a single ligament causes very little luxation. As successive ligaments were cut increasing displacement occured, but it was only after all the ligaments were cut

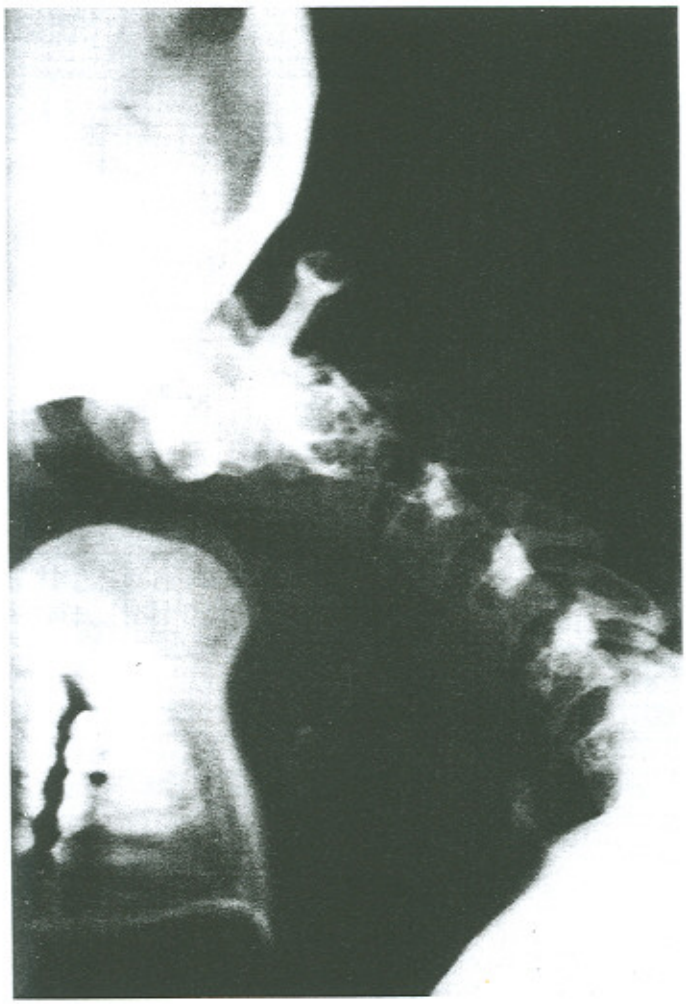

Figure 3: Flexion view shows forward displacement.

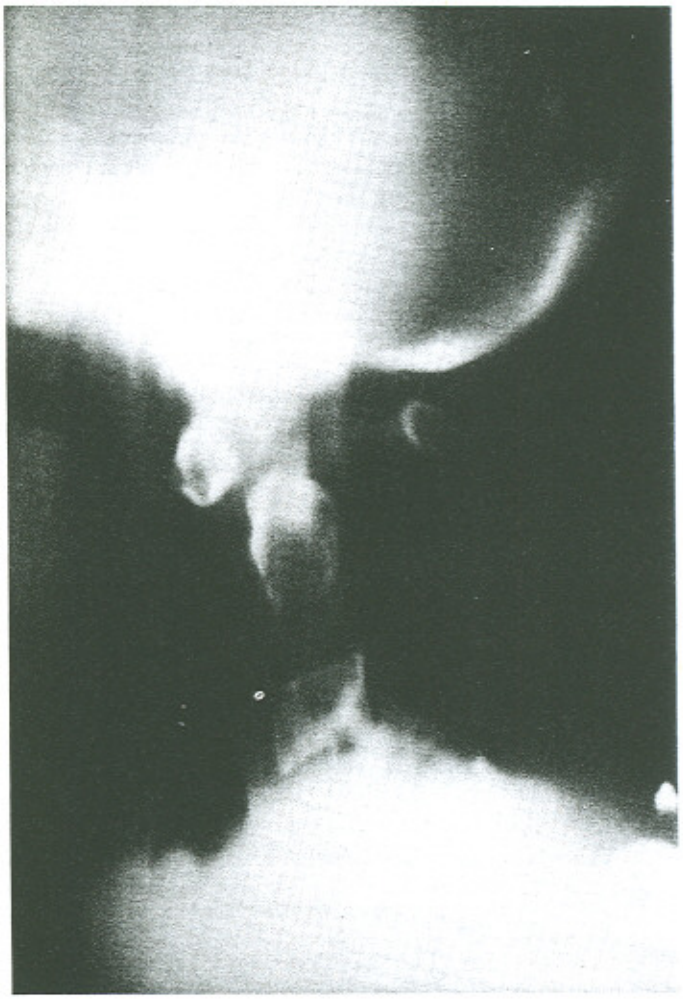

Figure 4: Tomogram shows absence of odontoid peg.

that catastrophic luxations were observed.

Failure of development during the early blastimal stage (approximately the sixth week of embryonic development) may result in a partial or complete agenesis of any vertebral element. Hirsh[11] feels that the failure of development of one ossific centre or failure of two centres to join together may be responsible for absent or separate bones. Deformity from a congenital anomaly may increase as the individual develops. When an agenesis is present, the subsequent weakness is compensated for by anomalous structures or by the development of increased strength in adjacent vertebrae. Agenesis or hypoplasia of varying degree is a common condition and may be encountered at any level of the spine, from an absent odontoid to an absent sacrum. Congenital abnormalities of the odontoid process can be divided into three basic types: agenesis, hypoplasia (partial absence) and non-fusion of the odontoid process. Fusion of the base of the odontoid does not occur radiographically until the age of twelve years and should not be confused with a fracture. Greenberg[12], however, has described five types of odontoid anomalies. Type 1 is the failure of the whole body of the odontoid to join the body of the axis and leads to an os odontoideum. Type 2 is the failure of the apical segment to join the body of the odontoid (ossiculum terminale). If the apical portion of the odontoid ossifies, but the odontoid body does not develop then agenesis of the odontoid base is present (Type 3). Type 4 is the failure of the apical segment of the odontoid to ossify and is responsible for agenesis of the apical segment. Type 5 is failure of the odontoid to ossify and may be responsible for agenesis of the odontoid.

Anomalies are prone to occur at the transition levels such as the atlanto-occipital, cervico-dorsal, dorso-lumbar or lumbosacral areas. The ligamentum flavum and fibrous structures are found to be unusually tough in the presence of bony deficiencies.

Treatment

This patient was treated successfully on three separate occasions when he had recurrences of neck pain and on one occasion when he experienced occipital headaches. Only in the original instance was there any known traumatic etiology, the remaining three episodes were without known causes. In all four episodes, the symptoms cleared up within three to four visits. In between episodes the patient was completely asymptomatic. In each case, treatment was very specific and was confined to the cervical spine below C2. A minimum of force was used and the head and upper cervical spine was stabilized as much as possible during the manipulative procedures.

Conclusion

Should patients with this defect have cervical spine fusion? Gillman[13] suggested that the defect alone should not be the sole criterion for a fusion. However, patients who have transitory or progressive neurological symptoms, with or without trauma, are candidates for fusion. In the event that surgical intervention is not instituted, these patients should be apprised of the various risks and should be warned to avoid all activities which might traumatize the head or neck, especially activities that involve contact sports.

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