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A case report of a congenital cleft of the anterior atlas arch: a rare variant of the atlas mimicking fracture

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Congenital anterior midline clefts of the atlas are rare developmental anomalies with only a few reported cases in the literature. This normal variant of the atlas results when the anterior arch fails to fuse during the ossification process. Series of cadaveric dissections have demonstrated anterior midline atlas clefts in 0.1–0.2% of the general population. Histological examinations have revealed that the bony defect, which ranges from 1–5 millimetres in width, is bridged by fibrocartilagenous tissue, resulting in what is generally believed to be a stable atlas.

Radiographically, congenital anterior clefts mimic many types of atlantal fractures, including Jefferson burst fractures, and vertical fractures of the anterior arches. The distinct radiographic appearances of congenital clefts and acute fractures of the atlas help to distinguish them from each other.

In the majority of cases, anterior clefts of the atlas remain undetected and have no clinical ramifications. Detection usually occurs during emergency post-traumatic radiographic imaging of the upper cervical spine. Under these circumstances, the presence of this congenital anomaly can make it difficult to differentiate between an acute fracture and a congenital variant. Where there is suspicion of fracture, computed tomography (CT) must be considered, as it is the most useful means of differentiating between these two clinical entities.

Chiropractors considering spinal manipulative therapy should be concerned with stability of an atlas

Les fentes congénitales antérieures médianes de l'atlas sont des anomalies de développement rares. Quelques cas seulement ont été signalés dans la littérature. Cette variante normale de l'atlas apparaît lorsque l'arc antérieur ne parvient pas à fusionner lors du processus d'ossification. Plusieurs séries de dissections de cadavres ont démontré des fentes antérieures médianes de l'atlas chez 0,1 à 0,2 % de la population globale. Des examens histologiques ont montré que le défaut osseux, allant de 1 à 5 mm de largeur, était couvert pas le tissu fibrocartilagineux, et ressemblait donc à un atlas stable.

Sur les radiographies, les fentes congénitales antérieures ressemblent à de nombreux types de fractures atloïdiennes, y compris les fractures de Jefferson et les fractures verticales des arcs antérieurs. La physionomie distincte des fentes congénitales et des fractures aiguës de l'atlas sur une radiographie permet de les distinguer les unes des autres.

Dans la majorité des cas, les fentes antérieures de l'atlas ne sont pas détectées et n'ont aucune ramification clinique. Elles sont généralement identifiées sur les radios d'urgence de la colonne cervicale supérieure qui sont effectuées à l'issue d'un traumatisme. Dans ces circonstances, la présence de cette anomalie congénitale peut compliquer le processus de différentiation entre une fracture aiguë et une variante congénitale.

Lorsqu'on suspecte une fracture, on doit envisager la tomographie par ordinateur, car il s'agit du moyen le plus utile de différencier ces deux entités cliniques.

Les chiropraticiens qui envisagent une manipulation

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with an anterior cleft, particularly in patients with a recent history of injury to the cervical spine. Appropriate clinical and radiographic examinations must be used to rule out cervical spine instability, before treatment is commenced. A reasonable course of treatment may include the judicious use of spinal manipulation.
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KEY WORDS: abnormalities; athletic injuries; atlas; cervical vertebrae; chiropractic; diagnosis, differential; radiography; sports medicine; manipulation.

vertébrale doivent s'inquiéter de la stabilité de l'atlas avec fente antérieure, particulièrement chez les patients présentant des antécédents récents de blessure de la colonne cervicale. Des examens cliniques et radiographiques appropriés doivent être utilisés pour écarter le risque d'instabilité de la colonne cervicale, avant de commencer le traitement. Un traitement raisonnable pourrait inclure des manipulations judicieuses de la colonne.
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MOTS CLÉS : anormalités ; blessures sportives ; atlas ; vertèbre cervicale ; chiropratique ; diagnostic ; différentiel ; radiographie ; médecine sportive ; manipulation.

Introduction

A cleft located on the anterior arch of the atlas represents a rare and typically inconsequential congenital anomaly which usually remains undetected. It is most commonly discovered incidentally, during emergency post-traumatic imaging of the upper cervical spine.[1,2,3,4] Under emergency situations, the presence of this congenital defect may complicate an individual's clinical presentation, as it can easily be mistaken for an acute fracture of the upper cervical spine.

Case report

A 19-year-old male presented to a chiropractic office with an acute neck injury sustained three days earlier during a high school football game. Immediately after tackling an opponent by ramming the player with the top of his helmet, the patient developed neck pain and was unable to complete the game. That evening he found it increasingly difficult to move his neck, but was otherwise symptom-free. He had no prior history of neck injury.

During examination, the patient held his neck rigidly and was in obvious pain. Cervical active ranges of motion were moderately decreased, with acute pain elicited by left lateral flexion, and extension. Passive extension combined with rotation and axial compression of the head was also painful. Pain was reported with palpation of the sternocleidomastoid, scalenus and trapezius muscles, and over the posterior joints of the C1 and C4 vertebral segments. Neurological examination including cranial nerve testing and fundoscopic examination was normal.

Plain-film radiographs of the cervical spine demonstrated a 2 millimetre widening of the lateral masses of the atlas (Figure 1). A vertical lucency was noted projecting over the odontoid process on the AP open mouth view. On the lateral view, the anterior tubercle of the atlas appeared hazy and hypertophic (Figure 2). Flexion/extension studies showed the C1–C2 joint to be stable. A CT scan of the upper cervical spine was obtained and showed no evidence of recent bony injury. A well corticated bony defect was noted in the anterior arch of the atlas, representing spondyloschisis (non-union) of the anterior arch (Figure 3).

The patient underwent a course of treatment which included ice, rest, electrotherapy, soft tissue therapy, spinal manipulation to the mid-cervical spine, and exercise. A consultation with an orthopaedic specialist followed six weeks after the injury. He was allowed to resume playing football immediately after the consultation, on condition that he remain pain-free and that repeat flexion/extension radiographs were negative for delayed instability. The patient played the remainder of the season without pain, complaining only of mild neck stiffness in the upper cervical spine which eventually resolved.

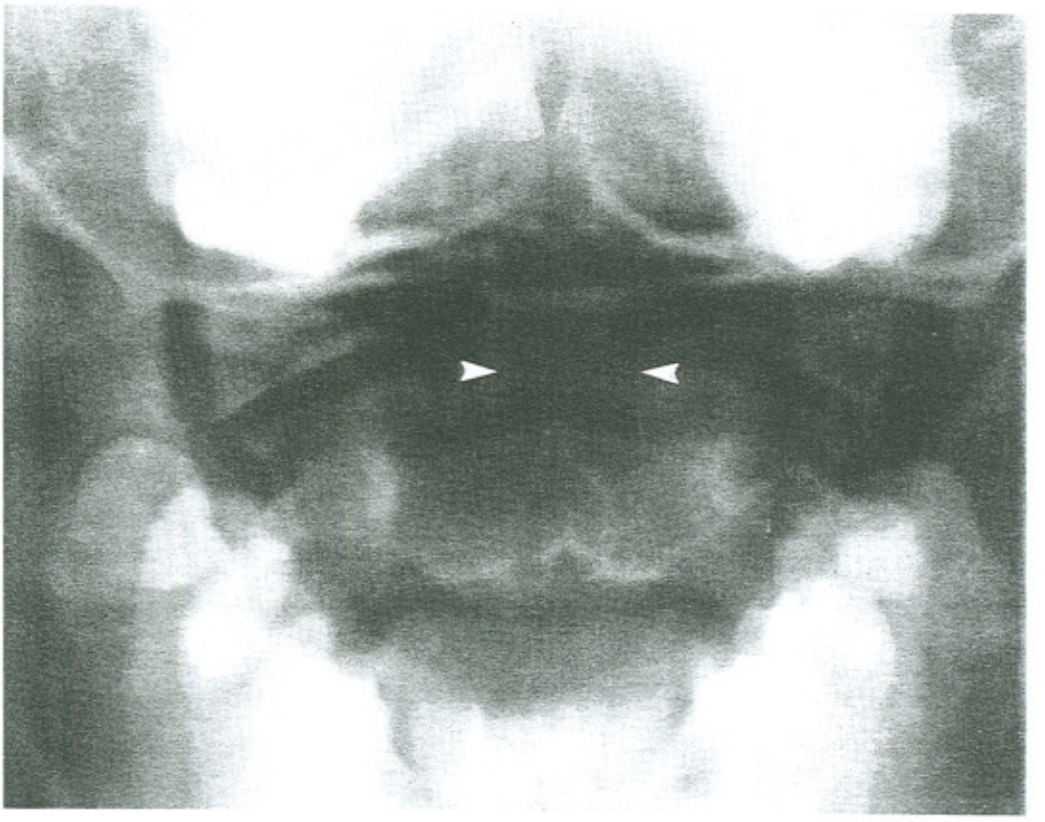

Figure 1 An AP open mouth view taken of a 19 year old male three days after sustaining a football injury to his cervical spine. A 2 millimetre widening of the lateral masses of the atlas and a vertical lucency projecting over the odontoid process is noted on this view. (arrow heads)

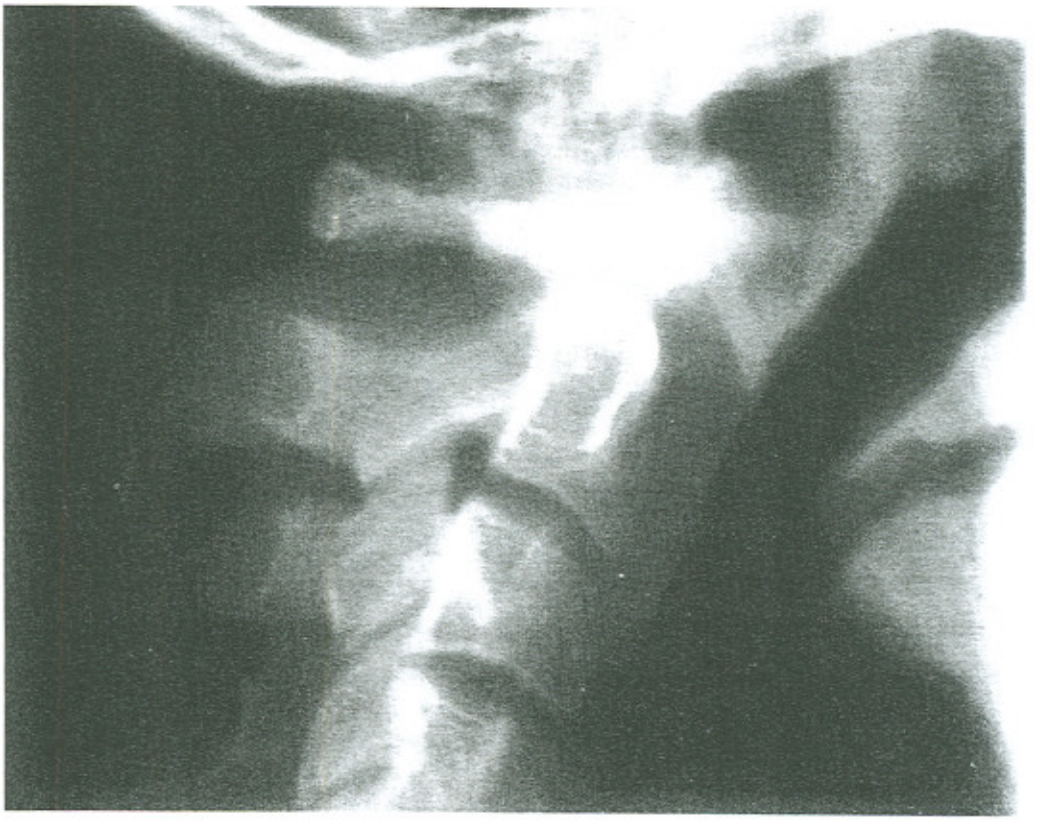

Figure 2 Lateral view of the cervical spine demonstrating the typical hazy and hypertrophic appearance of the anterior tubercle of the atlas in cases of congenital anterior arch clefts.

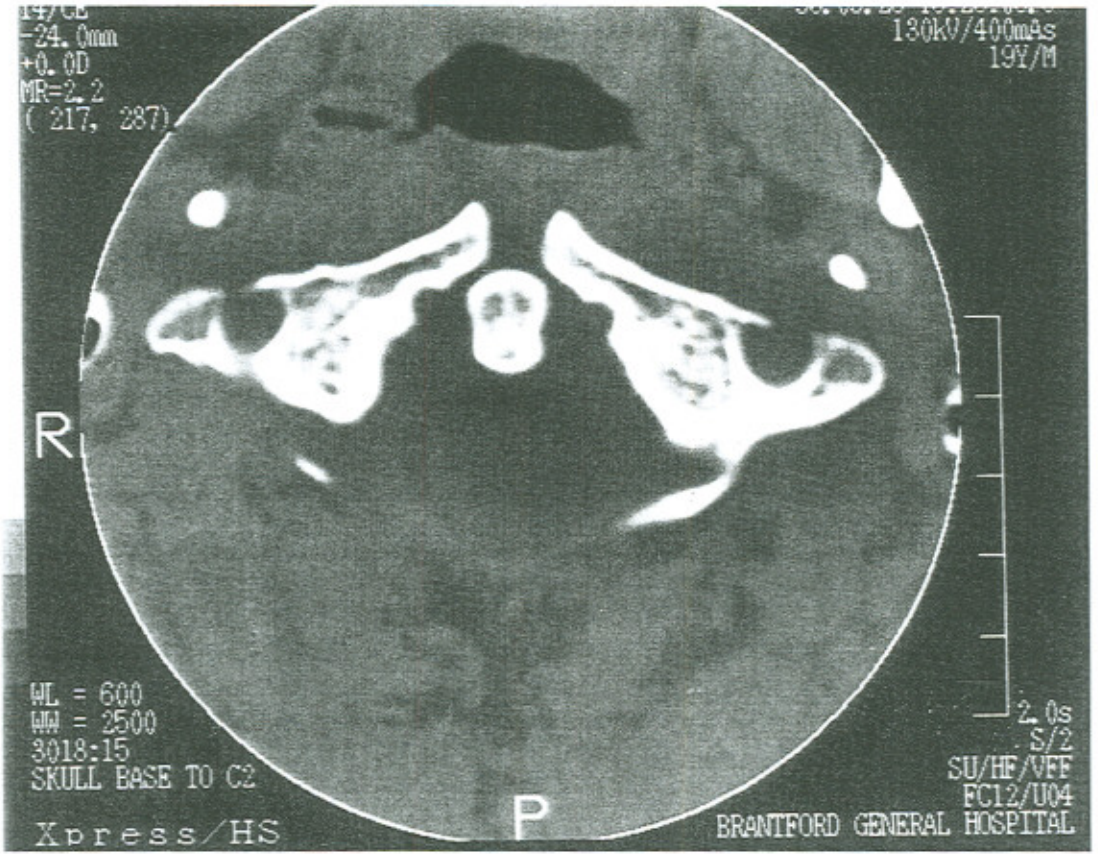

Figure 3 CT scan demonstrating a congenital cleft of the anterior arch of the atlas.

Discussion

Ossification of the Atlas

The atlas typically ossifies from three ossification centres (Figure 4). A centre appears at each of the lateral masses which gradually extend into the posterior arch where they unite by the third or fourth year of life. Another centre appears at the anterior tubercle, extending laterally and fusing with the lateral masses by the fifth to ninth year.[5,6] This pattern of anterior arch ossification, from a single, midline centre extending laterally to join the lateral masses, is the most commonly observed pattern. In some cases, the anterior arch may also be formed by the forward extension and fusion of the centres for the lateral masses. The anterior arch may also be formed by twin ossification centres.[1,5]

Congenital midline clefts of the anterior arch occur in one of two ways. A cleft may remain when the twin ossification centres at the anterior arch fail to fuse together.[1,7] In some cases, the ossification centre for the anterior tubercle fails to develop, and the lateral masses do not fuse anteriorly, resulting in a persistent anterior cleft.[7,8]

Epidemiology

Congenital midline clefts of the anterior arch of the atlas, also referred to as anterior spondyloschisis of the atlas or anterior arch rachischisis, are rare developmental anomalies with only a few cases reported in the literature.[1,2,9,10] In a series of cadaveric dissections, Geipel examined 2,749 atlases and found the presence of an isolated cleft through the anterior arch in only 0.1% of the cadavers.[11] Congenital midline clefts of the posterior arch (posterior spondyloschisis of the atlas or posterior arch rachischisis) are more common, appearing in 4% of 1600 dissections by Schultze and Buurman[12] and 2,749 dissections by Geipel.[11] In cases of combined anterior and posterior midline atlas clefts (anteroposterior spondyloschisis of the atlas or anteroposterior rachischisis), the bipartite atlas is referred to as a "split atlas" or "atlas bipartita."[6]

Histology

Histological examination of atlas specimens with anterior arch defects by Geipel determined that the two portions of

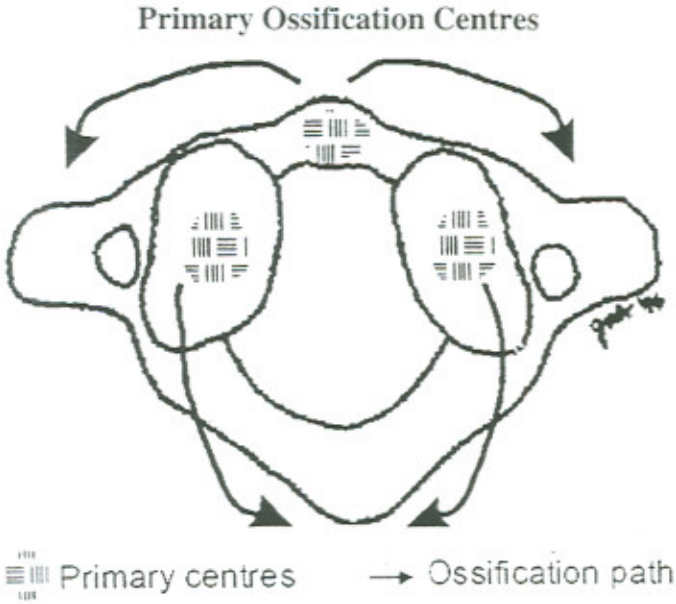

Figure 4 Location of primary ossification centres in the atlas and the direction of their extension during the process of ossification.

the arch were joined by fibrocartilagenous tissue.[11] The majority of anterior arch defects are midline and have a reported width ranging from 1–5 millimeters.[1]

Radiographic appearance and differential diagnosis

The characteristic midline lucency of anterior spondyloschisis, overlying the anterior arch of the atlas or the dens of the axis on AP open mouth, mimics some of the more common types of fractures observed in the atlas, including the Jefferson burst fracture and vertical fractures of the anterior and posterior arches.[6] Anterior arch clefts also mimic rare vertical fractures of the odontoid process of the axis. Jefferson burst fractures, which account for one-third of atlas fractures, result from an axial force applied to the head and neck, leading to disruption of the ring-like structure of the atlas, usually at the junction of the arches and the lateral masses.[2,4] Less often, an axial force to the head and neck can result in vertical fractures of the anterior and posterior arches of the atlas.

The characteristic features of a Jefferson burst fracture help to distinguish it from a congenital anterior atlas cleft (Table 1). A sharp, non-corticated vertical lucency viewed on AP open-mouth in association with asymmetry of the spaces between the atlas lateral masses and the dens of the axis is suggestive of fracture. Lateral spread of the lateral masses also suggests fracture of the atlas. In adults with a lateral spread of 3 millimeters or greater, a Jefferson burst fracture must be suspected. Lateral spread of 7 millimeters or greater most likely indicates complete rupture of the transverse ligament of the atlas, resulting in instability.[9] On the lateral view, anterior soft tissue swelling in the upper cervical region suggests a retropharyngeal hematoma, supporting the diagnosis of acute fracture.

Conversely, a midline vertical lucency overlying the arch of the atlas whose margins are smooth, well-corticated, and sclerotic, with lack of displacement of the lateral masses and adjacent soft tissue swelling, suggests a congenital cleft. A vertical lucency overlying the dens may simulate a vertical or longitudinal fracture of the dens but this type of fracture is so rare that it should raise the suspicion of a congenital cleft. While lateral spread of the atlas is generally considered to result from a fracture, it has been described in cases of combined anterior and posterior arch defects.[9] Combined anterior and posterior arch clefts may be associated with lateral widening of 1–2 millimeters, but this does not indicate abnormal spread of the atlas. Since these cases of congenital widening were not accompanied with clinical or radiographic signs of cervical instability they were judged to have no clinical ramifications.[1,3,7,8,13] We were unable to identify previously reported cases of lateral widening associated with isolated clefts of the anterior arch, as demonstrated in the patient discussed in this case report.

On a lateral radiograph, a normal anterior atlas arch appears crescenteric or half-moon in shape with surrounding cortical bone defining the anterior aspect of the atlantoaxial interspace.[9,14] In the presence of an anterior cleft, the anterior arch is visibly plump and rounded due to chronic stresses resulting from altered biomechanics. The anterior arch appears to overlap the dens, making it impossible to evaluate the atlantoaxial interspace. The arch may also appear to have double anterior margins.[14]

In cases of patients presenting with a history of trauma to the upper cervical spine, it is often difficult to differentiate radiographically between an acute fracture and a congenital variant.[2,3,9] Computed tomography (CT) is the most useful means to differentiate between congenital clefts and fractures of the atlas and it must be considered in a patient with a history of trauma where clinical and radiographic findings are suggestive of fracture.[1,6,14] Findings

Table 1
Radiographic Appearance of Atlas Anterior Cleft vs Burst Fracture

	ANTERIOR CLEFT	BURST FRACTURE
Xray View APOM	smooth, corticated, and sclerotic lucency, usually located in the midline	sharp, non-corticated lucency at the junction between the lateral masses and the arches
	usually not associated with lateral widening of the lateral masses	associated with lateral widening of the lateral masses
	if lateral widening present, not greater than 3 mm	lateral widening of 3 mm or greater
Lateral	hazy, rounded and hypertrophic anterior arch	the anterior arch is crescenteric in shape and well outlined by cortical bone
	evaluation of the atlanto-axial interval difficult due to apparent anterior arch overlap	the atlanto-axial interval is easily evaluated
	no adjacent soft tissue swelling	anterior soft tissue swelling

consistent with an anterior atlas cleft seen on axial CT include a midline anterior arch defect ranging from 1–5 millimeters in width with smooth, sharply defined, well-corticated margins. This is in contrast to the sharp, irregular, and non-corticated appearance of an acute fracture.[1] Additionally, there will be no evidence of soft tissue swelling adjacent to the bony defects and no abnormal orientation between the atlas and the axis.[1,3]

Clinical implications

Congenital variations in the cervical anatomy generally have no clinical ramifications except when associated with trauma or other conditions that lead to instability of the cervical spine.[13] While a congenital cleft of the anterior arch presumably results in a weaker atlas, it is believed to be of little consequence.[8,13] Geipel's confirmation by histologic examination of specimens that the two portions of the anterior arch of the atlas are joined by fibrocartilage has led most authors to believe that the vertebra is stable. Some authors believe that wide congenital clefts, joined by fibrous tissue only, may be associated with atlas instability, though it is not clear which measurement in millimetres was considered to represent a wide cleft.[14] An increased risk of fracture may also be associated with congenital clefts, as suggested by a number of reported cases, but these may only represent an incidental association.[1,6,13]

For the chiropractor, the stability of the atlas with an anterior spondyloschisis is important when considering spinal manipulation as a treatment. We have not been able to find previous studies that address this issue in the scientific literature. The existing research on anterior spondyloschisis consists solely of case reports and case series and their conclusions may have limited generalizability to general practice. We were also unable to find studies which directly examined the strength or stability of the atlas in cases of anterior spondyloschisis. Consequently when considering spinal manipulation in cases of anterior spondyloschisis, clinical judgement must prevail. The chiropractor must safeguard against the possibility of cer

vical spinal instability by clinical examination, radiographic evaluation, and when indicated, specialist consultation. Once cervical instability has been ruled out, spinal manipulation, used judiciously, may reasonably be selected for inclusion in a course of treatment.

Conclusion

An anterior arch cleft of the atlas may mimic many types of atlantal fractures, making radiographic diagnosis of this congenital variant difficult. Recognition of this normal variant is further complicated by the post-traumatic setting where it is most commonly discovered.

The patient discussed in this report is a case in point. Post-traumatic radiographic examination of the patient's upper cervical spine revealed hallmark signs of burst fractures of the atlas, including widening of the lateral masses and a vertical lucency overlying the odontoid. A CT scan helped differentiate between a fracture and a congenital anomaly, which allowed for appropriate management strategies to be undertaken.

Since strength and stability in anterior arch midline clefts have not been conclusively studied, the chiropractor must exercise caution when considering spinal manipulation in patients with this normal variant. Once fracture and instability have been ruled out, the judicious use of spinal manipulation may be reasonably included in the treatment of upper cervical spine injuries.

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