Os odontoideum with associated multidirectional atlantoaxial instability: imaging and clinical considerations

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Acquired anomalies of the craniovertebral junction although rare, may result in serious clinical sequella. The prevalence of craniovertebral anomalies remains unknown since they may remain clinically silent. Os odontoideum is one such anomaly. The etiology remains controversial over whether it is a post-traumatic or congenital condition but the clinical considerations of os odontoideum remain the same. Chiropractors may encounter os odontoideum in the subclinical patient with the potential for deleterious effects with manipulation. Radiography is essential to its detection.

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Introduction
The purpose of this paper is to demonstrate the multidirectional upper cervical instability present in an individual with os odontoideum but without significant complaints. Patients may present with symptoms ranging from mild local neck pain to myelopathy and vascular compromise.1–12 The variability of signs and symptoms may make diagnosis difficult and it is often discovered incidently with routine cervical radiographs.10,13 Atlantoaxial instability secondary to os odontoideum is a serious condition that may require surgical stabilization.1,14 A case of os odontoideum with associated multidirectional instability is presented with discussion focused on diagnostic imaging and clinical implications.

Case report
A 45-year-old female presented with right cervicothoracic pain and occipital headaches. The headaches were of
recent onset, without apparent cause, and had been con-
stant the week prior to her initial presentation. She re-
ported a feeling of “clunking” in her neck.

She had a previous history of “fainting spells” with
three distinct episodes approximately 5 years previously;
in one case requiring stitches to the back of her head. At
that time she was evaluated by a neurologist, but she was
not treated and she did not recall a diagnosis being made.
She reported many injuries related to horseback riding in-
cluding a left femoral fracture in her 20’s requiring open
reduction and internal fixation. She had been struck by a
car on her left side at the age of 10 but did not recall any
significant resulting injuries.

Examination findings
The patient walked with a normal gait and her Rhomberg
test was normal. Motor and reflex testing was normal and
symmetric in the upper and lower extremities. Bilateral
plantar responses were down-going. No sensory deficits
were elicited with regard to tactile stimulation (both
sharp and dull sensation tested) in the upper and lower
extremities. Proprioception and vibratory sensation was
intact and screening evaluation of the cranial nerves (III-
XII) and Valsalva maneuver were unremarkable. Active
ranges of motion of the cervical spine were full and pain
free in flexion, extension, bilateral rotation and lateral
flexion. Passive cervical flexion exhibited a palpable
clicking in the upper cervical spine and she complained
of tenderness in the upper cervical area during this
maneuver. At that time, the decision was made to proceed
with radiographic examination before more provocative
orthopaedic testing was performed.

Radiological examination
Cervical radiographs were performed due to the recent
onset of cervical and occipital pain, past history of trau-
ma and palpable clicking detected during passive cervical
flexion. Anteroposterior open mouth and lower cervical,
lateral neutral cervical and flexion-extension stress views
were performed. As the patient had been able to perform
bilateral cervical lateral flexion during active range of
motion testing without complaint, and there was no his-
torical complaint associated with this movement, addi-
tional A-P open mouth views were taken in lateral flexion
after review of the neutral open mouth film. The neutral
lateral cervical film (Figure 1) reveals the hypertrophied
anterior tubercle of C1 in the approximate position nor-
mally occupied by the odontoid process. A prominent re-
versal of the cervical lordosis and extensive degenerative
changes in the lower cervical spine are evident. Cervical
extension (Figure 2) and flexion (Figure 3) radiographs
demonstrate atlantoaxial instability in the sagittal plane.
An Instability Index of 35.7 % was calculated.

The distance from the posterior aspect of the ossicle to
the spinolaminar junction line of C2 with the cervical
spine in extension (represented as D_{ext}) is measured at
24 mm. The ossicle is difficult to visualize on the lateral
projection (arrows), but the 14 mm sagittal plane transla-
tion of the sclerotic and hypertrophied anterior tubercle
of the atlas is apparent. The space available for the cord
(SAC), being the minimum distance from the posterior
aspect of the odontoid or axis to the nearest posterior

![Figure 1](image1.png)

Figure 1  Lateral cervical radiograph in neutral. The
hypertrophied anterior tubercle of the atlas is visualized in
the position normally reserved by the dens (arrows). Note the
prominent reversal of the cervical lordosis.
structure (foramen magnum or posterior ring of the atlas), is measured at 18 mm.

The posterior aspect of the anterior tubercle exhibits a convex configuration which has been described as a “molding” defect. The inferior aspect of the C1 posterior arch also exhibits a concave configuration. The AP open mouth projection in neutral position (Figure 4) reveals an irregular blunt tooth peg (type II) at the superior aspect of the C2 vertebral body where it should be connected to the odontoid process and the atlantoaxial joint is offset (arrows). Active right (Figure 5) and left (Figure 6) lateral flexion open mouth radiographs demonstrate 13 mm of coronal plane translation of the lateral masses of the atlas in relation to the axis. 6,16

Computed tomography and magnetic resonance imaging
Coronal reformatted CT demonstrates an oval well-corticated ossicle which appears to have maintained an orthotopic relationship with the body of C2. The odontoid base has an asymmetrical orientation (Figure 7). A sagittal reformatted CT image obtained in the neutral position demonstrates that the ossicle is in fact dystopic or displaced and appears to have migrated superiorly to the clivus (Figure 8). A sagittal T2-weighted MR image demonstrates the dystopic relationship of the ossicle with mild effacement of the adjacent anterior cord as well as posteriorly by the C2 spinous process (Figure 9).
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Figure 4  Anteroposterior open-mouth view in the neutral position demonstrates 3 mm offset (arrows) of the lateral masses of the atlas in relation to the axis.

Figure 5  Anteroposterior open-mouth view during active right lateral flexion demonstrates the coronal plane instability in this patient.

Figure 6  Anteroposterior open-mouth view during active left lateral flexion. The total coronal plane translation of C1 relative to C2 is measured at 13 mm.

Figure 7  Coronal reformatted CT demonstrates discontinuity between the oval well-corticated odontoid process and the asymmetrical dens base of C2.
Diagnosis
Os odontoideum with associated multidirectional instability of C1 on C2. The principal differential diagnosis of os odontoideum is an acute fracture of the odontoid process. Owing to the characteristically smooth cortex of the ossicle and the adjacent C2 vertebral body peg, the absence of a recent history of acute trauma, and the sclerosis and hypertrophy of the anterior tubercle of the atlas, we excluded the diagnosis of acute odontoid fracture.

Discussion
Acquired anomalies of the odontoid process are uncommon\textsuperscript{10,17} and detection may occur incidentally on cervical radiographs.\textsuperscript{10,13,17} The etiology of os odontoideum remains controversial and debate continues over whether it is a post-traumatic or congenital condition.\textsuperscript{1,5–7,15,16,18,19–23} Regardless of the origin, the clinical management remains the same.\textsuperscript{7} Clinically, patients may present with many puzzling symptoms ranging from local pain and occipital neuralgia to myelopathy and vertebrobasilar ischemia.\textsuperscript{1–4,6–12} Relative lack of symptoms in most cases has been attributed to the abundant space within the spinal canal at the C1–2 level. This anatomic arrangement in which free space allows for a degree of pathological variation has been described as Steel’s Rule of Thirds.\textsuperscript{6,11} As there is poor correlation between the amount of instability and the presence of clinical signs, the validity of manual upper-cervical stability testing is questionable\textsuperscript{24} and functional radiography is the standard.

Due to the high incidence of atlanto-axial instability in cases of os odontoideum, cervical flexion-extension radiographs should be obtained to document the degree of instability.\textsuperscript{20,24–26} Our case demonstrates instability in both sagittal and coronal translation\textsuperscript{27} associated with a deficient odontoid. Hypertrophy and sclerosis of the anterior tubercle of the atlas has been used to differentiate os odontoideum from acute fracture of the dens.\textsuperscript{19} Hypertrophy of the anterior arch of atlas and disruption of the spinolaminar junction line are radiographic signs associated with, but not specific for, os odontoideum.\textsuperscript{19,28} Remodelling of the inferior surface of the posterior arch of atlas is also visualized in this case, and this variation is likely the result of similar longstanding stress due to C2 spinous contact. As our patient was asymptomatic during active cervical range of motion and with consideration of the established sagittal translation, open-mouth lateral flexion views were performed to determine the degree of cervical instability.
multidirectional instability at C1–C2. The asymmetrical irregularity of the odontoid peg visualized on the anteroposterior open mouth projection suggests an acquired etiology. This case has been described as a dystopic blunt-tooth type.29

CT and MR imaging have greatly advanced our understanding of os odontoideum and its relationship to neurovascular compromise. The arch-peg ratio as measured on sagittal CT scan has been utilized23 in an attempt to quantify the hypertrophy of the anterior arch due to chronic stress of the unstable atlanto-axial junction. Our case exhibits this elevation of the arch-peg ratio consistent with longstanding instability (Figure 8). CT imaging also clearly illustrates the discontinuity of the dens with the C2 vertebral body as well as superior-migration of the ossicle toward the clivus. The proposed mechanism for the superior movement is contracture of the unopposed alar ligaments.1,7,17 MR images were obtained in a neutral position and do not demonstrate the extremes of sagittal translation that may indicate more significant cord compression.30

The associated segmental instability and compromise of the space available for the spinal cord (SAC) can lead to transient or progressive myelopathy.3,5,6,10 A SAC of 13 mm or less has been identified as a critical measure related to potential myelopathy.9,10 The presented case demonstrates a SAC measurement exceeding 13 mm.

Shirasaki et al.27 found the degree of instability was not as significant a factor due to morphological variation in SAC.19 They hypothesized that the development of cervical myelopathy was related to the distance from the posterior aspect of the ossicle to the spinolaminar junction of the axis (Dext). They found a Dext value of 16 mm or less correlated with dynamic canal stenosis causing myelopathy related to posterior translation. The case presented in this report had a Dext value exceeding the 16 mm threshold at the time of investigation.

Another utilized indicator of atlantoaxial instability is the Instability Index measuring the dynamic sagittal diameter of the spinal canal: It is calculated as (maximum diameter – minimum diameter)/maximum diameter x 100.8,9 The maximum anteroposterior diameter of the spinal canal was measured at 28 mm, and the minimum anteroposterior diameter of the spinal canal was measured at 18 mm in our case. This yields an index of 35.7% consistent with severe atlanto-axial dislocation. An Instability Index of greater than 40% was found to increase the probability of cord signs.8

Watanabe et al.9 identified the limitation of utilizing the sagittal plane Instability Index in the evaluation of cases of os odontoideum with multidirectional instability. It is further suggested that progressive myelopathy may result from a repetitive type insult to the cord due to the excessive excursion of the atlas on axis causing meningeal hypertrophy,12 edema and atrophy of the spinal cord.9

Another concern due to the related pathomechanics of os odontoideum is potential vertebral artery hemodynamic compromise.10 The tortuous path of the vertebral arteries in the most mobile area of the spine, further unchecked by an unstable os odontoideum, leaves the arteries vulnerable to direct compression as well as repetitive injury which can lead to intimal damage.2,8 Numerous vascular symptoms have been associated with this mechanism including vestibular4 and cerebellar disturbances.2,5 Magnetic resonance angiography has been used to determine compromise of the vertebral arteries due to excessive atlanto-axial instability.5,31

Os odontoideum, whether the result of congenital osseous malformation or prior unrecognized trauma, requires the same radiologic and clinical workup. Surgical stabilization of the upper cervical spine has been proposed for progressive symptomatology including intractable neck pain, transient or progressive myelopathy, vertebral artery insufficiency and worsening instability.10,20

In this case presentation, the patient was advised of her condition, precautions to observe (i.e. restrict participation in sports and high risk activities) and referred for neurosurgical consultation. At two years after the diagnosis and identification of instability, she remained symptom-free and comfortable with monitoring of her condition. She chose not to undergo a prophylactic surgical procedure.

Conclusion
This case demonstrates significant multidirectional upper cervical instability associated with os odontoideum. Identification of os odontoideum is essential to chiropractors and others who practice manual therapy. The condition represents an absolute contraindication for upper cervical manipulation. The potential of this entity to remain clini-
cally silent emphasizes the necessity of appropriate eval-
uation including radiographs and advanced imaging.

References