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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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KEY WORDS: Os odontoideum, atlantoaxial instability, manipulation.

Quoique peu fréquentes, les anomalies acquises de la jonction bulbo-médullaire peuvent laisser des séquelles cliniques graves. Le taux de prévalence des anomalies bulbo-médullaires est inconnu, la recherche clinique ne s'étant pas encore penchée sur la question. L'os odontoïde fait partie de ce type d'anomalies. Le diagnostic visant à établir s'il s'agit d'un état post-traumatique ou congénital reste controversé mais les observations cliniques concernant cette anomalie demeurent les mêmes. Les chiropraticiens peuvent rencontrer des cas d'os odontoïde chez les patients subcliniques et cette anomalie est susceptible d'avoir un effet néfaste sur la santé en cas de manipulation. Les radiographies sont essentielles pour établir un diagnostic.
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MOTS CLÉS : Os odontoïde, instabilité atlantoaxiale, manipulation.

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

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recent onset, without apparent cause, and had been constant the week prior to her initial presentation. She reported 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 including 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 trauma 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 historical complaint associated with this movement, additional 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 normally occupied by the odontoid process. A prominent reversal 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 translation 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 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.

Figure 2 Lateral cervical radiograph in extension demonstrates os odontoideum and associated instability at C1C2. Note the posterior sagittal translation of the hypertrophied anterior tubercle of C1 with extension.

Figure 3 Lateral cervical radiograph in flexion demonstrates the anterior sagittal translation of the hypertrophied anterior tubercle relative to C2.

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.[15] 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).

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.

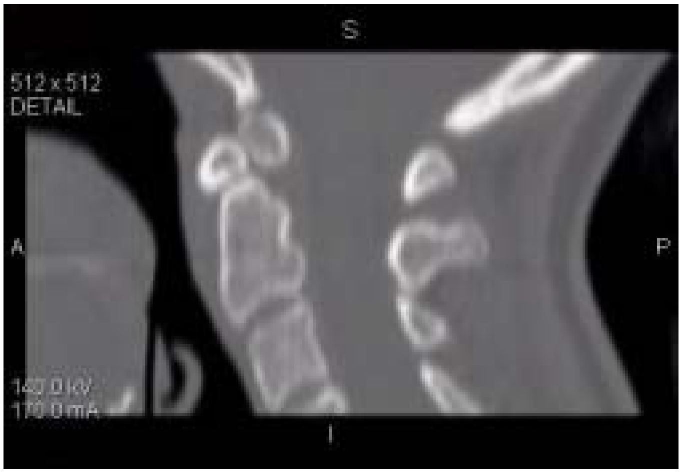

Figure 8 Sagittal reformatted CT image demonstrates the posteriorly displaced ossicle.

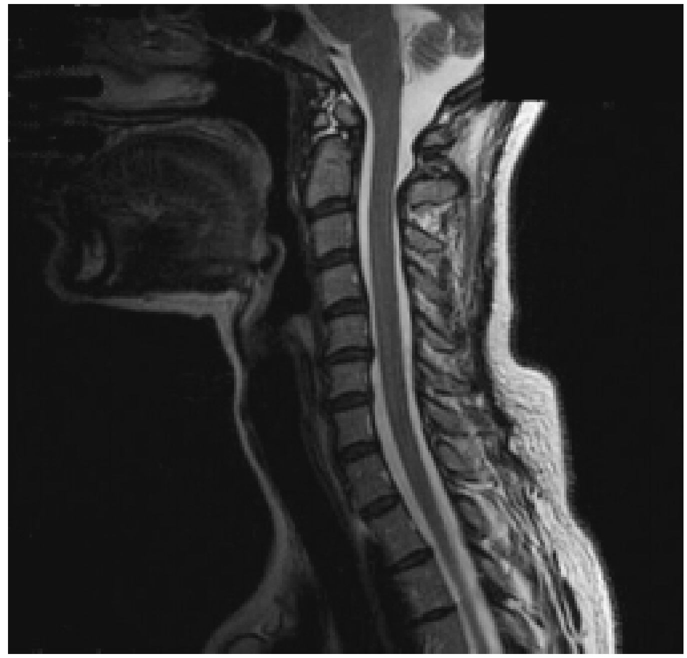

Figure 9 A sagittal T2-weighted MR image demonstrates the posterior displacement of the ossicle with mild effacement of the adjacent anterior cord as well as posteriorly by the C2 spinous process.

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[10,17] and detection may occur incidentally on cervical radiographs.[10,13,17] The etiology of os odontoideum remains controversial and debate continues over whether it is a post-traumatic or congenital condition.[1,5–7,15,16,18,19–23] Regardless of the origin, the clinical management remains the same.[7] Clinically, patients may present with many puzzling symptoms ranging from local pain and occipital neuralgia to myelopathy and vertebrobasilar ischemia.[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.[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[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.[20,24–26] Our case demonstrates instability in both sagittal and coronal translation[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.[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.[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 multidirectional instability at C1–C2. The asymmetrical irregularity of the odontoid peg visualized on the antero-posterior 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 utilized[23] 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 (D_{ext}). They found a D_{ext} 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 D_{ext} 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 vestibular[4] 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 clinically silent emphasizes the necessity of appropriate evaluation including radiographs and advanced imaging.

References

1 Dai L, Yuan W, Ni B, Jia L. Os odontoideum: Etiology, diagnosis and management. Surg Neurol 2000; 53:106–109.
2 Sasaki H, Itoh T, Takei H, Hayashi M. Os odontoideum with cerebellar infarction. Spine 2000; 25(9):1178–1181.
3 Chang H, Park J, Kim K, Choi W. Retro-dental reactive lesions related to development of myelopathy in patients with atlantoaxial instability secondary to os odontoideum. Spine 2000; 25(21):2777–2783.
4 Galli J, Tartaglione, Calo L, Ottaviani F. Os odontoideum in a patient with cervical vertigo: A case report. Am J of Otolaryngology 2001; 22(5):371–373.
5 Fukuda M, Aiba T, Akiyama K, Nishiyama K, Ozawa T. Cerebellar infarction secondary to os odontoideum. Journal of Clinical Neuroscience 2003; 10(5):625–627.
6 Ramos LS, Taylor JAM, Lackey G, Lindsay D. Os odontoideum with sagittal and coronal plane instability: A report of three cases. Topics In Diagnostic Radiology and Advanced Imaging 1996; 3(1):5–12.
7 Clements W, Mezue W, Mathew. Os odontoideum - congenital or acquired? – that's not the question. Injury 1995; 26(9):640–642.
8 Takakuwa T, Hiroi S, Hasegawa H, Hurukawa K, Endo S, Shimamura T. Os odontoideum with vertebral artery occlusion. Spine 1994; 19(4):460–2.
9 Watanabe M, Toyama Y, Fujimura Y. Atlantoaxial instability in os odontoideum with myelopathy. Spine 1996; 21(12):1435–1439.
10 Hensinger R. Osseous anomalies of the craniovertebral junction. Spine 1986; 11(4):323–333.
11 Ebraheim N, Lu M, Yang H. The effect of translation of the C1-C2 on the spinal canal. Clinical Orthopaedics and Related Research 1998; 351:222–229.
12 Thurlow R. Odontoid agenesis with atlanto-axial luxation. JCCA 1986; 30(2):77–80.
13 Crnec M, Cassidy D, Mierau D, Yong-Hing K. Os odontoideum: A case report. JCCA 1986; 30(2):83–85.
14 Hosono N, Yonenobu K, Ebara S, Ono K. Cineradiographic motion analysis of atlantoaxial instability in os odontoideum. Spine 1991; 16(10):S480-S482.
15 Marchiori, D. Clinical imaging. St. Louis: Mosby, 1999:406–09.
16 Taylor JAM, Resnick D. Skeletal imaging: atlas of the spine and extremities. Philadelphia: W. B. Saunders Company, 2000:55–6.
17 Fielding W, Hensinger R, Hawkins, R. Os odontoideum. J Bone Jt Surg 1980; 62-A(3):376–383.
18 Stevens J, Chong W, Barber C, Kendall B, Crockard H. A new appraisal of abnormalities of the odontoid process associated with atlanto-axial subluxation and neurological disability. Brain 1994; 117:133–148.
19 Vickers E. Atlantoaxial anomalies with particular emphasis on os odontoideum. J Manipulative Physiol Therap 1990; 13(8):471–476.
20 Hughes T, Richman J, Rothfus W. Diagnosis of os odontoideum using kinematic magnetic resonance imaging. Spine 1999; 24(7):715–718.
21 Verska J, Anderson P. Os odontoideum : A case report of one identical twin. Spine 1997; 22(6):706–709.
22 Matricali G, Matricali B. Os odontoideum: Radiological, anatomical and histological examination of a post-mortem specimen. J Orthopaed Traumatol 2000; 1:57–661.
23 Fagan A, Askin G, Earwaker. The jigsaw sign. A reliable indicator of congenital aetiology in os odontoideum. Eur Spine J 2004; 13:295–300.
24 Swinkels R, Oostendorp R. Upper cervical instability: Fact or fiction. J Manipulative Physiol Therap 1996; 19(3):185–193.
25 McGregor M, Mior S, Shannon H, Hagino C, Schut B. The clinical usefulness of flexion-extension radiographs in the cervical spine. Top Clin Chiro 1995; 2(3):19–28.
26 Yi-Kai L, Yun-Kun Z, Shi-Zhen Z. Diagnostic value on signs of cervical vertebrae with radiological examination. J Manipulative Physiol Therap 1998; 21(9):617–620.
27 Shirasaki N, Okada K, Oka S, Hosono N, Yonenobu K, Ono K. Os odontoideum with posterior atlantoaxial instability. Spine 1991; 16(7):706–715.
28 Holt G, Helms C, Munk P, Gillespy T. Hypertrophy of C-1 anterior arch: Useful sign to distinguish os odontoideum from acute dens fracture. Radiology 1989; 173:207–209.
29 Matsui H, Imada K, Tsuji H. Radiographic classification of os odontoideum and Its clinical significance. Spine 1997; 22(15):1706–1709.
30 Laiho K, Soini I, Kautiainen H, Kauppi M. Can we rely on magnetic resonance imaging when evaluating unstable atlantoaxial subluxation? Ann Rheum Dis 2003; 62:254–256.
31 Roche C, O'Malley M, Dorgan J, Carty H. A pictorial review of atlanto-axial rotatory fixation: Key points for the radiologist. Clinical Radiology 2001; 56:947–958.