Case Report

Spontaneous regression of retro-odontoid post traumatic cicatrix following occipitocervical fixation

ABSTRACT

This case report describes a patient who presented with myelopathy secondary to a large retro-odontoid post traumatic cicatrix. The objective of this study was to discuss the clinical presentation, pathogenesis, imaging, and surgical management of pseudoarthrosis tissue mass associated with odontoid nonunion. Atlantoaxial subluxation (AAS) has been widely reported in patients with rheumatoid arthritis. AAS leads to repeated cycles of partial tear and repair of ligaments around the altantoaxial complex, resulting in the formation of periodontoid mass (pseudotumor). It is thought that formation of retro-odontoid post traumatic mass (cicatrix), in certain cases of odontoid fracture, is because of similar pathology. This is a retrospective review of case note. Here, the patient underwent posterior decompression through a C1–C2 laminectomy and occipitocervical (C0–C4) fusion with instrumentation, which resulted in dramatic improvement in his symptoms and spontaneous regression of retro-odontoid post traumatic cicatrix. We have described an interesting and a rare case of a large pseudoarthrosis tissue mass associated with odontoid nonunion, which regressed following stand-alone posterior instrumentation. To the best of our knowledge, only a handful of such cases of spontaneous regression of retro-odontoid post traumatic cicatrix following occipitocervical fixation have been described in literature, and our case adds to the growing list of such cases and may help in understanding the natural history of the disease process one day. Although rare, post traumatic cicatrix should be considered as a differential diagnosis of enhancing retro-odontoid mass, especially if there is any history of cervical spine trauma.

Keywords: Occipitocervical fixation, post traumatic cicatrix, retro-odontoid

INTRODUCTION

Odontoid fracture constitutes about 10% of all cervical spine fractures. However optimum management of type II odontoid fracture especially in the elderly population is still unclear. The natural history of untreated odontoid fracture is also unknown. Some patients tend to remain asymptomatic whereas others can present with myelopathy weeks to years after the initial injury. We have presented challenging case of a patient with a large retro-odontoid post traumatic mass who was treated with stand-alone posterior instrumentation and discussed aetiology, pathogenesis and management of this clinical entity.

CASE REPORT

A 79-year-old Caucasian male presented to our unit with a 3-month history of worsening gait, progressive weakness of limbs, and clumsiness of his hands. He was from a well-to-do family and a nonsmoker; he consumed occasional alcohol and denied any substance misuse. He also denied any recent weight loss, cough, fever, foreign travel, or bladder/bowel dysfunction. On examination, he was apyrexial, Glasgow coma scale was 15, and cranial nerves were grossly intact. However, he was quadriparetic (power 2/5) with increased tone in all the four limbs. He also had brisk reflexes

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throughout, positive Hoffman's sign, sustained clonus, and upgoing plantars.

Magnetic resonance imaging (MRI) of his brain was normal. MRI of his cervical spine showed a retro-odontoid mass, which appeared isointense on T1-weighted sequences and hypointense on T2-weighted sequences [Figure 1a-c]. There was evidence of anterior subluxation of C2 on C1 with severe cord compression at that level, anteriorly from the mass and posteriorly from the posterior arch of atlas with signal change in the cord. Postgadolinium administration, there was inhomogeneous enhancement of the mass along with peripheral enhancement of the capsule [Figure 1d]. Computed tomography (CT) scan of the cervical spine confirmed the presence of a large retro-odontoid mass [Figure 1e]. The scans were reviewed by the consultant neuroradiologist, and differential diagnosis was (a) pannus, (b) metastasis, (c) chordoma, (d) myeloma, (e) and lymphoma. There was no clinical evidence of rheumatoid arthritis, psoriasis, or gout. C-reactive protein, erythrocyte sedimentation rate, serum calcium, and uric acid were normal. Rheumatoid factor and protein electrophoresis were negative too, and Bence–Jones protein was absent. CT scans of his chest, abdomen, and pelvis did not reveal any abnormality. The patient was reassessed and a detailed history was taken during which the patient admitted having a trivial neck injury 4 years ago. Although he had not attended a hospital for this, he has been having persistent neck pain following this episode. The scans were re-reviewed by the neuroradiologist and odontoid nonunion with a large pseudoarthrosis tissue mass (post traumatic cicatrix) was added to the differential diagnosis. After extensive discussion with the patient and his family and given his age, performance status, and comorbidities, we decided to do a stand-alone posterior cervical procedure. The two main aims of the treatment were decompression of neural structures and stabilization of atlantoaxial subluxation (AAS) which led to the formation of the pseudotumor in the first place. We decided to perform decompression in addition to posterior cervical fusion because of the presence of severe cord compression at the affected level and rapidly progressive history of neurological deterioration in 3 months' time. We recognized the fact that stand-alone posterior cervical fusion without decompression would have led to regression of retro-odontoid mass, however we were unsure of the duration of this process. As the patient had rapidly progressive symptoms, we combined fusion with decompression to immediately relieve pressure from the spinal cord.

He underwent posterior cervical decompression through a C1–C2 laminectomy and occipitocervical (C0–C4) fusion with instrumentation. The patient was positioned prone with neck in flexion and the position was secured with the help of a head clamp. A midline skin incision was made from inion to C5 spinous process, and a subperiosteal exposure of the occiput and cervical spine was carried out. A high-speed drill was used to normalize the bony anatomy of the lateral masses. Occipital plates and lateral mass screws into C3 and C4 were then placed. C1–C2 laminectomy was performed, and craniovertebral junction was decompressed. Neck of the patient was then repositioned and rods were secured to occipital plate and cervical screws. Decortication was performed at the joints and artificial bone graft was placed around it to aid in fusion. Postoperative period was uneventful. When he was seen in clinic in 3 months' time, he had power 4/5 in all the four limbs and he was mobilizing with the help of a frame. His manual dexterity had also markedly improved. Repeat MRI done 4 months' postprocedure showed

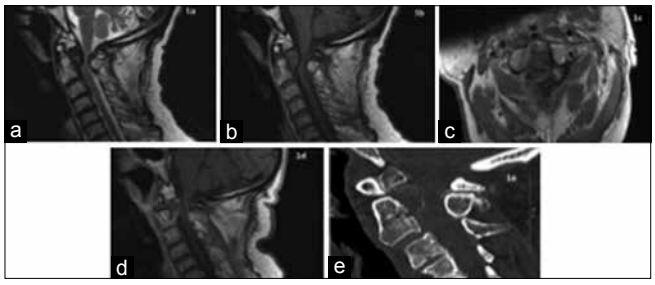

Figure 1: (a) T2-weighted sagittal magnetic resonance imaging of cervical spine showing hypointense retro-odontoid mass causing cord compression. (b) T1-weighted sagittal magnetic resonance imaging of cervical spine showing isointense retro-odontoid mass causing cord compression. (c) T2-weighted axial magnetic resonance imaging of cervical spine showing severe cord compression at the level of retro-odontoid mass. (d) Postgadolinium administration, T1-weighted sagittal magnetic resonance imaging showing inhomogeneous enhancement of the retro-odontoid mass along with peripheral enhancement of the capsule. (e) Computed tomography scan of the cervical spine showing a large retro-odontoid mass

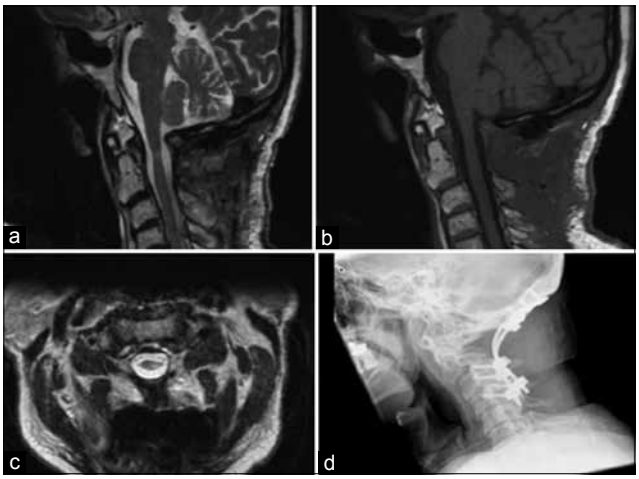

Figure 2: (a) T2-weighted sagittal magnetic resonance imaging of cervical spine showing disappearance of the retro-odontoid mass following occipitocervical fixation. (b) T1-weighted sagittal magnetic resonance imaging of cervical spine showing disappearance of the retro-odontoid mass following occipitocervical fixation. (c) T2-weighted axial magnetic resonance imaging of cervical spine showing decompressed spinal cord following disappearance of the retro-odontoid mass. (d) Lateral cervical spine X-ray showing occipitocervical fixation

spontaneous regression of the retro-odontoid mass [Figure 2a-d]. One year following the surgery, he was independently mobile and there was no evidence of any tumor or inflammatory disease. However, because of lack of histopathological diagnosis, our diagnosis of odontoid nonunion with a large pseudoarthrosis tissue mass represents a diagnosis of exclusion.

DISCUSSION

AAS in patients with rheumatoid arthritis has been extensively described in literature, and though it is mostly asymptomatic, it can cause cord compression and lead to cervical myelopathy.[1] Similarly, formation of soft tissue mass at the craniovertebral junction secondary to chronic AAS is a well-defined clinical phenomenon.[2,3] Sze *et al*. described this mass as "periodontoid pseudotumor."[3] It is most commonly seen as pannus in rheumatoid arthritis,[4] and the standard treatment of such periodontoid mass used to be transoral resection of the mass along with posterior stabilization.[5] However, anterior spinal surgery was associated with a number of complications, and following the reports of spontaneous disappearance of the mass following posterior stabilization only,[2,4] nowadays, stand-alone posterior cervical fixation is usually performed for rheumatoid pannus.[6,7] Other conditions causing chronic AAS such as psoriasis,[8] diffuse idiopathic skeletal hyperostosis,[9] and os odontoideum[10] can also produce such mass at the craniovertebral junction, and there have been reports of spontaneous resolution of the mass in psoriasis[8] and os odontoideum[10] following stand-alone posterior cervical fixation. It is thought that hypertrophic pseudotumor in such cases is formed as a result of repeated cycles of partial tear and repair of ligaments around the altantoaxial complex.[11]

Odontoid fracture constitutes about 10% of all cervical spine fractures. Some patients who sustain odontoid fracture may not survive the insult whereas some others fail to attend the hospital because of lack of symptoms. Odontoid fractures were classified into three types by Anderson and D'Alonzo in 1974,[12] and the most common is type II fracture. Management of odontoid fracture still remains challenging and controversial, especially in the elderly population, as it is associated with a significant morbidity and mortality. Most surgeons recommend external orthosis for the management of type I and type III fractures. However, optimum management of type II odontoid fracture, especially in the elderly population, is unclear as some recommend external orthosis whereas others recommend internal fixation. There is also controversy regarding anterior or posterior approach to the cervical spine for the management of such fractures.[13] The natural history of untreated odontoid fracture is unknown. Some patients tend to remain asymptomatic whereas others can present with myelopathy weeks to years after the initial injury.[14] Our patient had sustained an injury to the neck 4 years ago, though symptoms of myelopathy appeared in the past 3 months only. In literature, there are only a handful of cases, which have described spontaneous regression of post traumatic cicatrix, following stand-alone posterior cervical fixation [Table 1]. Our case of delayed cervical myelopathy and spontaneous regression of symptomatic retro-odontoid mass following occipitocervical fixation adds to the growing list of such cases and may help in understanding the natural history of the disease process in the future. Our case highlights the fact that, though rare, retro-odontoid post traumatic cicatrix can cause diagnostic dilemma and should be considered as a differential diagnosis in a patient presenting with retro-odontoid mass. As retro-odontoid post traumatic cicatrix enhances with gadolinium, it can be confused with a malignant growth in a nonrheumatoid patient which can have grave clinical and psychological implications for the patient. Our case was challenging as authors[15] in the past have felt that, in cases with large retro-odontoid mass, stand-alone posterior instrumentation may not be adequate to decompress the spine. In retrospect, ideally, we should have performed C1–C2 fusion in this patient with C1 lateral mass screws and C2 pars screws with a fusion in the facet joints. We recognize that, by fusing the occiput to C1, we unnecessarily decreased 50% of this patient's flexion-extension motion, and by disrupting semispinalis cervicis muscle, whose major attachment is at C2, we have predisposed the patient to postoperative kyphosis. However, despite our extensive experience in C1–C2 fusion, at the time of presentation, we were reluctant to instrument C2. One of the initial differential diagnoses was of malignancy and we were reluctant to instrument a segment of the spine with potential malignant involvement, which could have resulted in failure of the instrumentation. We had considered using augmented screws; however, given the patient's age and his wishes to minimize any potential risk of postoperative construct failure, we decided to perform occipitocervical fusion. Our diagnosis of odontoid nonunion with a large pseudoarthrosis tissue mass is essentially a retrospective diagnosis in the absence of any histopathological sample. Ideally, we had also decided to biopsy the lesion as a first-step procedure at the time of occipitocervical fixation, however in view of the patient's age, comorbid conditions, and wishes, we chose to perform posterior cervical fixation with decompression.

Odontoid fractures have a high rate of nonunion. Hence, fractures which are not reported, like our patient, or even fractures which fail to unite with treatment or malunite may subsequently lead to AAS and myelopathy. Many surgeons

Table 1: Reported cases of stand-alone posterior cervical instrumentation for the management of post traumatic retro-odontoid mass

Author	Journal	Year	Patient's age	Treatment
Lansen *et al.*	Journal of Neurosurgery	1990	56	Transoral resection and posterior cervical fusion*
Pare *et al.*	Neurosurgery	1995	38	Posterior cervical fusion (C1–C3)
Young and Boyko	Journal of Clinical Neuroscience	2002	75, 65, and 52	C1–C2 fixation
Shah *et al.*	World Neurosurgery	2016	42 and 16	Atlantoaxial lateral facetal distraction and fixation
Meng and Liu	The Spine Journal	2016	50	Posterior reduction and occipitocervical fusion
Ikegami *et al.*	Spine	2015	52	Lateral atlantoaxial joint puncture and arthrography
Tobari *et al.*	Journal of Orthopaedic Surgery	2014	89 and 85	C1 laminectomy and posterior fixation from the occiput to C3 and rigid cervical collar, respectively
Ohnishi *et al.*	European Spine Journal	2015	70	Intradural cyst resection without fusion was performed through the anterolateral approach
Lin *et al.*	Indian Journal of Orthopaedics	2014	64	Laminectomy and C1–C2 fixation
Klineberg *et al.*	Evidence-Based Spine-Care Journal	2014	83	Transoral decompression under fluoroscopic visualization
Luksanapruksa *et al.*	Journal of the Medical Association of Thailand	2013	67	Occiput to C3 fusion with plating, posterior arch of atlas resection, transoral odontoidectomy, and mass removal
Kakutani *et al.*	European Spine Journal	2013	76, 64, 73, 87, 70, 82, and 77	C1 laminectomy, C1 laminectomy, C1 laminectomy + C3–C6 laminoplasty, C1 laminectomy, C1 laminectomy, C1 laminectomy + C3–C6 laminoplasty, C1 laminectomy + C3–C6 laminoplasty, respectively
Takeuchi *et al.*	The Spine Journal	2011	76	Decompression of the lesion combined with atlantoaxial fixation
Ogata *et al.*	Journal of Spinal Disorders and Techniques	2009	58	Posterior fixation between C1 and C3 without resection of the retro-odontoid mass
Yamazaki *et al.*	Journal of Clinical Neuroscience	2009	49	C1 dome-like laminectomy and posterior C1–C2 polyaxial screw–rod fixation
Takami *et al.*	Neurologia medico-chirurgica	2007	68	C1 laminectomy and posterior C1–C2 fixation with individual screw placement in C1 and C2
Sato *et al.*	Neurologia medico-chirurgica	2006	38	Decompressive laminectomy
Lin *et al.*	Spinal cord	2006	74	Decompression surgery was performed with a laminectomy at C3–C6 levels and removal of the ligamentum flavum extended to the posterior arch of the atlas
Isono *et al.*	Surgical neurology	2001	74	Decompression through a laminectomy of C1 and occipitocervical fixation
Hasegawa *et al.*	Neurologia medico-chirurgica	2000	86	C1 hemilaminectomy and upper partial hemilaminectomy of C2
Jun	Spine	1999	61	Posterior C1–C2 transarticular screw fixation
Takemoto *et al.*	Spine	2015	72, 69, 71, 75, 82, 84, 74, 79, 78, and 78	C1 laminectomy without fusion

*The authors believe that posterior cervical fusion resulted in the regression of the traumatic periodontoid hypertrophic cicatrix as immediate postoperative scan after transoral resection did not show significant reduction in the size of the cicatrix

recommend operative intervention for mobile nonunited odontoid fracture even in the absence of myelopathy;[14,15] however, there are also reports where mobile nonunited odontoid fracture, in the absence of myelopathy, have been managed conservatively.[16] We were reluctant to do flexion/extension X-rays of the cervical spine to assess the mobility of the nonunited fragments because of severe cord compromise and rapidly progressive symptoms. Some authors[17] feel that even fibrous nonunion of the odontoid fracture in the elderly population is acceptable in terms of function and that there is no significant risk of myelopathy[13] whereas others[18] consider a nonunion as an absolute indication for surgery. We feel that the decision to operate or not to operate should be on a case-to-case basis. Even though the natural history of untreated odontoid fracture is unknown and there are reports of nonunited odontoid fractures presenting with myelopathy years after the initial injury, the true incidence of odontoid fracture, especially in the elderly population, is unknown and it may be possible that there are a number of asymptomatic patients with odontoid fracture (some of which may be nonunited) in the general population, who may never present to a hospital. Similarly, when deciding not to operate, one must bear in mind the fact that even a trivial injury can have catastrophic consequences for some of these asymptomatic patients with nonunited odontoid fracture.

CONCLUSION

Management of symptomatic odontoid nonunion with a large pseudoarthrosis tissue mass should be on a case-to-case

basis, depending on clinical signs and symptoms, radiological findings, surgeon's expertise, and patient's comorbidities. Although rare, post traumatic cicatrix can cause dilemma in diagnosis and management, especially in the elderly population where the history of trauma may be trivial. Our case also highlights the fact that, because of the rarity of this condition (i) it is hardly ever mentioned by neuroradiologists in their differential diagnosis, (ii) most of our experience is based on case reports, and (iii) there is no standardized treatment and treatment has been based on experience derived from the management of rheumatoid pannus. Our case also highlights the importance of detailed history taking in patients presenting with retro-odontoid mass, as in some cases, the patient may be elderly and the history of trauma may be trivial. However, one should be careful in drawing conclusions from these isolated reports of regression of retro-odontoid post traumatic cicatrix following stand-alone posterior instrumentation, as there are reports of spontaneous regression of retro-odontoid cyst (whose etiopathogenesis is considered to be AAS too) with cervical collar alone.[19,20]

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Conflicts of interest

There are no conflicts of interest.

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