Oxford City Publishers

Case Reports in Orthopaedic Surgery Journal

Case Report | Vol 2 Iss 3

Purely Ligamentous Combined Atlanto-Occipital and Atlanto-Axial Dissociation

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Received: July 27, 2023; **Accepted:** August 18, 2023; **Published:** August 29, 2023

Abstract

Craniocervical dissociation is typically a fatal condition that occurs secondary to a high-energy trauma, most commonly MVA's, auto-pedestrian accidents, and falls from elevated heights. Disruption of the internal and external ligamentous stabilizers of the upper cervical spine predisposes to chronic neurologic sequelae in patients who survive. We report a rare case of purely ligamentous atlanto-occipital and atlantoaxial dissociation in a polytrauma patient after being struck by a vehicle as a pedestrian. We describe the unique clinical, imaging, intraoperative findings, and outcomes associated with this injury. Remarkable findings included absent sensorimotor function in all extremities, no rectal tone, autonomic dysregulation, as well as absent bulbocavernosus, babinski and clonus reflexes. Despite the poor prognosis associated with this injury, posterior cervical fusion from the occiput to C4 was indicated for care purposes and mitigation of additional neurologic complications. Moreover, craniocervical stabilization remains the gold standard for treatment of cervical instability when indicated, regardless of an anticipated good or poor prognosis.

Introduction

Craniocervical dissociation (CCD) can be regarded as a comprehensive term to describe atlanto-occipital dissociation (AOD) and atlantoaxial dissociation (AAD) [1]. Both result from disruption of internal and external ligaments that stabilize the craniocervical junction between the occiput and C2 vertebrae, often with osseous lesions. Due to the proximity of the brainstem and high-energy trauma typically associated with these injuries, mortality rates and risk of permanent neurologic sequelae are high. Cervical spine injuries have an average reported incidence of 49.7% as the etiology for fatality in blunt trauma with 20% due to CCD [2,3]. The most common mechanisms are motor vehicle accidents, auto-pedestrian accidents, and fall from above ground level. The resultant forces can lead to disruption of the ligamentous structures that stabilize the craniocervical junction (CCJ) [2-4].

We report a rare case of purely ligamentous CCD in which AOD, AAD, and high cervical quadriplegia occurred without concomitant osseous lesions. We describe the clinical findings, associated imaging, and postoperative outcomes.

Case Report

A 35-year-old male presented to our level one trauma center as a pedestrian struck by a high-speed motor vehicle with a glasgow coma scale score of 3T. Neurologic evaluation revealed spinal shock among a plethora of other injuries. Initial head/neck CT scan demonstrated a Powers Ratio of 1.1, an Atlanto-Dens Interval (ADI) of 10.4, and a space available for the cord (SAC) of 9.2 (Figure 1). MRI confirmed complete ligamentous disruption of the atlanto-occipital and atlanto-axial joint (Figure 2). The exam remained unchanged following return of the bulbocavernosus reflex, thus classifying the patient as ASIA A distal to the occiput on the ASIA Impairment Scale.

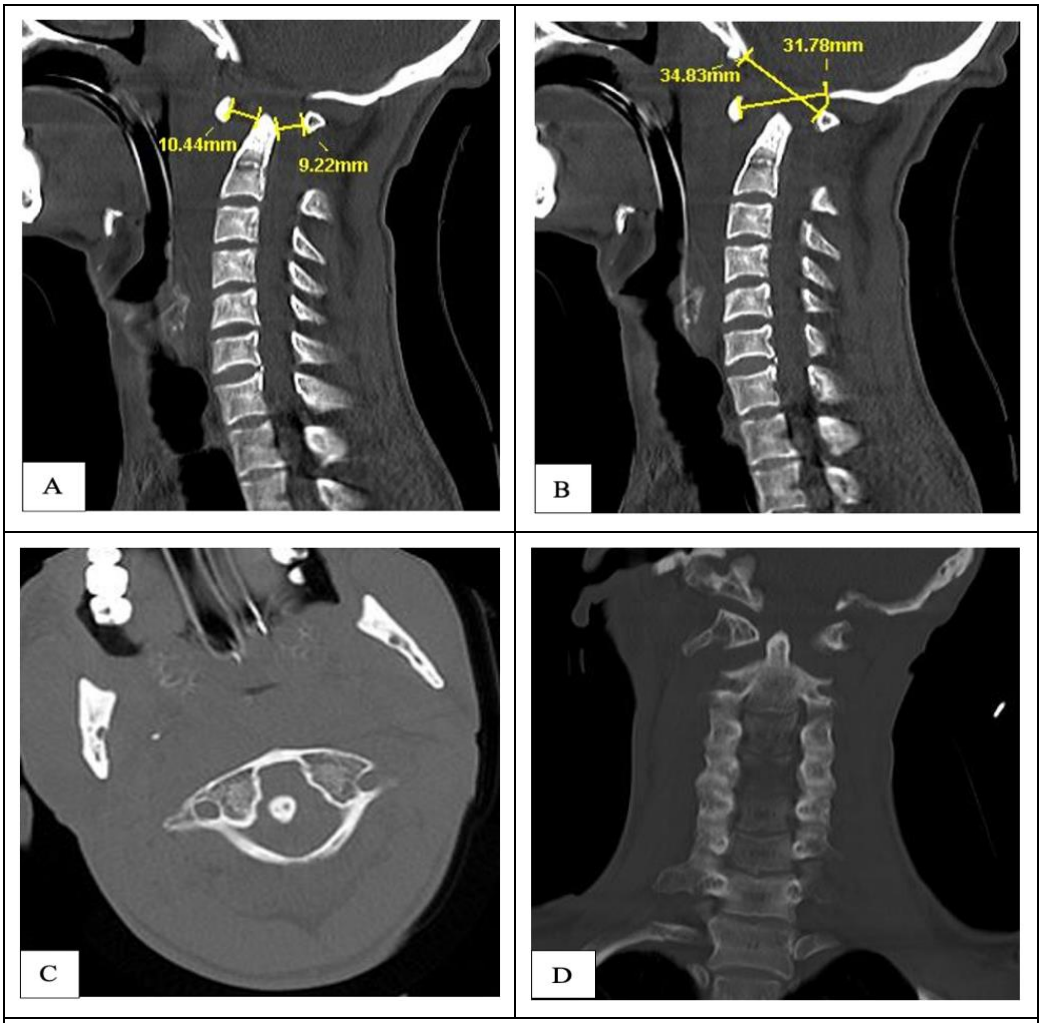

Figure 1(A): Sagittal CT scan demonstrating Atlanto-Dens Interval (ADI) of 10.4, Space available for the cord (SAC) of 9.2, and **(B):** Powers Ratio of 1.1. **(C):** Axial and **(D):** Coronal CT scan demonstrating incongruent atlanto-axial joint.

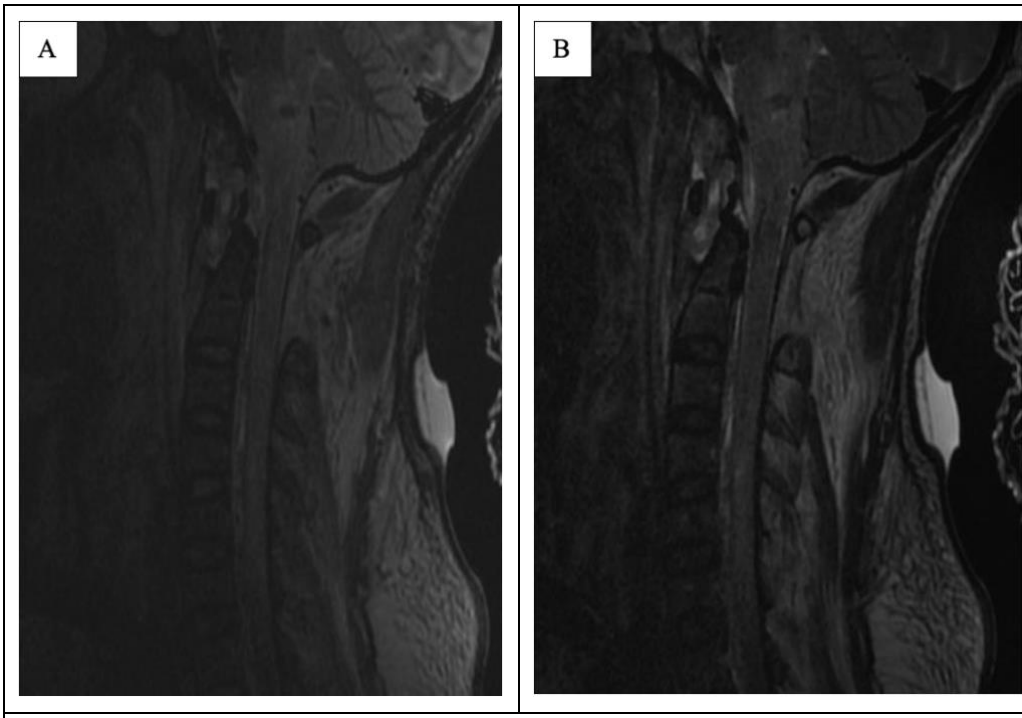

Figure 2(A): STIR and **(B):** T2-weighted sagittal MRI scans demonstrating complete ligamentous disruption of the atlanto-occipital and atlanto-axial joint.

Posterior cervical fusion was indicated for care purposes and mitigation of additional neurologic complications. Pre- and post-positional neuromonitoring demonstrated no motor or sensory evoked potentials. The spine was stabilized via posterior cervical fusion from the occiput to C4 utilizing a suboccipital plate, C1 lateral mass screws, C2 pars screws, and C3-4 lateral mass screws. Reduction was performed with gentle posterior traction and maintained with C1-2 sublaminar wires and an appropriately sized rod (Figure 3). Motor or sensory evoked potentials remained unchanged throughout the case.

Postoperatively, the patient was managed in the surgical intensive care unit for one month prior to discharge to an extended care nursing facility. His postoperative period was riddled with complications associated with his polytraumatic injury including acute kidney injury requiring hemodialysis. The patient was lost to follow-up, however records showed he passed away two months later due to ongoing complications from his polytraumatic injury.

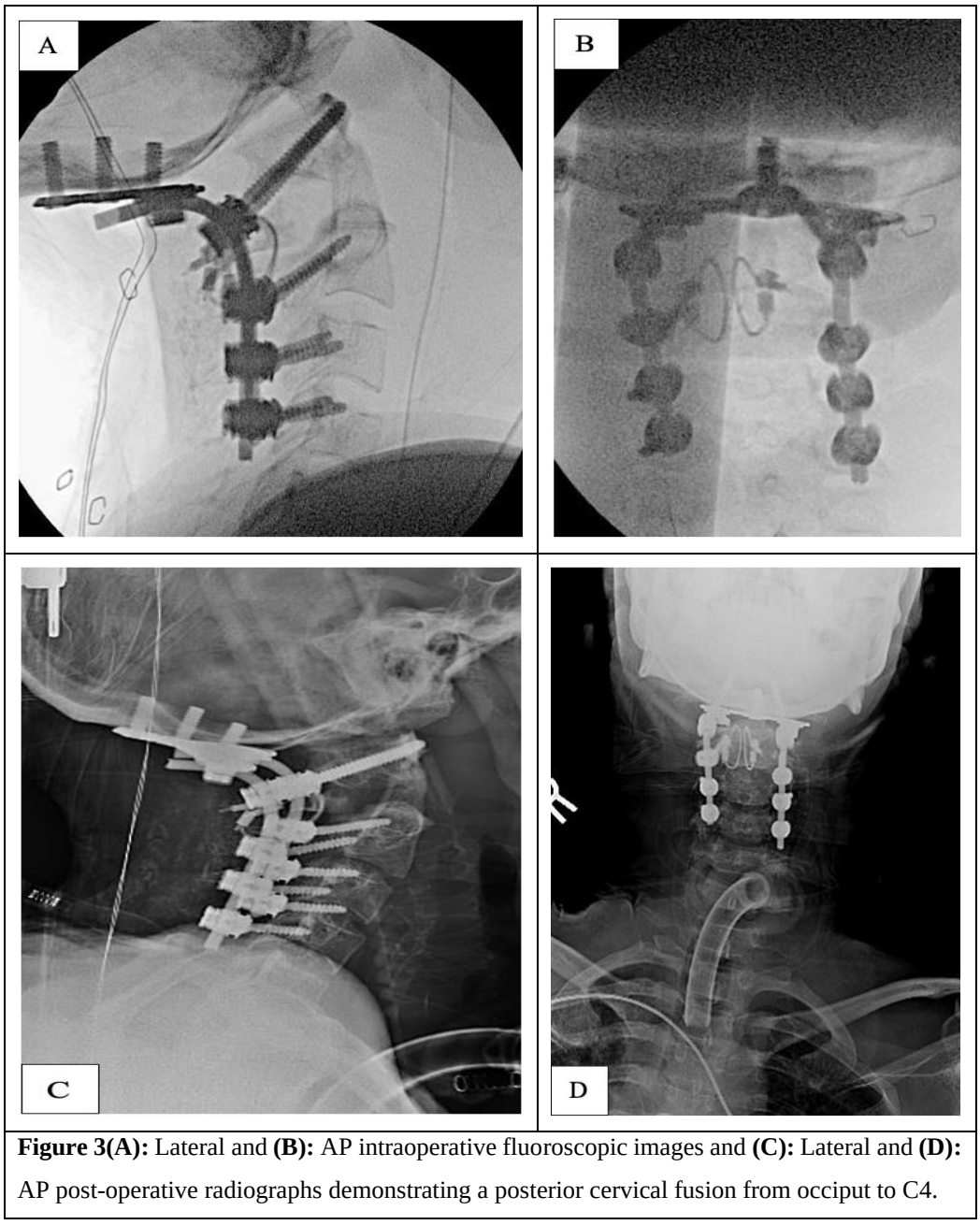

Figure 3(A): Lateral and **(B):** AP intraoperative fluoroscopic images and **(C):** Lateral and **(D):** AP post-operative radiographs demonstrating a posterior cervical fusion from occiput to C4.

Discussion

We report a purely ligamentous combined atlanto-occipital and atlanto-axial dissociation in a polytrauma patient with a GCS of 3T, sensorimotor deficits, and hemodynamic instability. Once stable, the patient underwent occiput-C4 posterior cervical fusion to minimize neurologic sequelae and for care purposes. While several studies have described AOD and AAD occurring in isolation, there is limited literature reporting these injuries concomitantly. Of the reports available, such patients were reported to have a concomitant cervical fracture or few to no other complications [4-7]. In a retrospective review by Cooper et al of 69 patients with CCD, the seven patients who survived to discharge had a Glasgow Coma Scale greater than 3, were hemodynamically stable, and did not require resuscitative measures [1].

Motor vehicle accidents, auto-pedestrian accidents, and falls from above ground level are the three most common high-energy mechanisms predisposing to CCD [1,5,6,8]. A retrospective study by Tepper et al showed no protection from AOD despite being restrained, sober, or wearing a helmet in a motorcycle accident when compared to all traffic deaths [9]. Children under the age of eleven have a greater predilection for CCD due to larger relative head size, hypoplastic occipital condyles, general ligamentous laxity, and less obliquely oriented atlanto-occipital joints [2,6]. Chronic inflammatory conditions such ankylosing spondylitis and rheumatoid arthritis, in addition to genetic diseases such as Morquio or Klippel-Feil syndrome have all been described as non-traumatic etiologies [6,10].

Stability to the CCJ is attributed to the complex ligamentous network that encompasses the occiput, atlas, and axis [2,11]. While the tectorial membrane resists cervical hyperextension, the transverse and paired alar ligaments, components of the cruciate ligament, resist hyperflexion and anterolateral flexion, respectively. [2,8,11]. Ligamentous disruption between the occiput and C2, with or without osseous lesions, result in AOD and AAD [8]. High-energy traumatic forces can result in disruption by any amalgam of shearing, rotational, distractive, translational, hyperflexive, or hyperextensive forces [1,2,4]. As such, presentation of CCD can range anywhere from asymptomatic to fatal [5]. Although cervical spine injuries with ensuing neurologic sequelae are the most common associated complications, thoracoabdominal and additional fractures are common in this polytrauma cohort.

Improvements in field resuscitation and stabilization with a rigid cervical collar, as well as diagnostic protocols utilizing multidetector computed tomography (MDCT), has led to greater awareness for high-suspicion patients despite the condition being typically fatal [2]. An autopsy study by Bucholz and Burkhead estimated a 1% chance of survival from AOD alone; extrapolating this to concurrent AAD diminishes the chances of survival even further [5,6,12,13]. Cervical management must be meticulous as loss of ligamentous integrity can lead to over distraction from even simple cranial traction [2,8,12]. In patients with severe CCD, halo vest constructs can be applied after manual fluoroscopic guided reduction for provisionally stabilization [8]. Halo vest immobilization alone is typically not sufficient to control occipital-cervical injuries and prevent neurologic injury; craniocervical fixation with modern segmental screw-based constructs improves neurologic outcomes and lowers patient complications [4,8].

In patients with a delayed diagnosis, 40% developed profound neurological deficits by hospital day two [2]. The two major prognostic factors are time to diagnosis and cervical stabilization with initial workup often limited to imaging [2,4]. Indiscernible landmarks and cases of low-clinical suspicion can result in missed diagnoses when using lateral cervical radiographs alone [1,2]. MDCT is the gold standard for diagnosis of CCD [2]. Suggestive findings include Powers ratio >1.0, basion-dens interval (normal 8.3 +/- 4.2mm), occipital condyle-C1 interval (normal 0.89 ± 0.12mm), lateral mass interval (normal <3.5mm), occiput-C1 distance (normal <5mm), C1-C2/C2-C3 ratio (normal <2.5), space available for cord (normal >19mm), as well as atlantoaxial (>1.2mm) and occipitocervical (>1.0mm) joint intervals [1,2,5-7,14,15]. Abnormality of the basion-dens interval was reported as the most common finding in CCD followed by an aberrant C1-C2/C2-C3 ratio, Powers ratio, and occipital condyle-C1 interval [15]. MRI can evaluate soft tissue injury in those whose neurologic status cannot be evaluated by 48 hours of injury; in turn, dynamic fluoroscopy can be used for an unclear diagnosis [2,6,11,15].

Conclusion

We present a rare case of purely ligamentous CCD managed with occiput-C4 posterior cervical fusion for care purposes and mitigation of additional neurologic complications. CCD is often a result of high energy trauma with a poor prognosis. Occipitocervical stabilization remains the gold standard for treatment for this inherently unstable pathology.

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