

An Unusual AAD with Basilar Invagination – Problems of Wrong Surgery Re-exploration

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ABSTRACT

Atlanto axial dislocations are rare yet devastating to the patients unfortunate enough to have them. AADs result in symptoms ranging from respiratory depression and CO₂ narcosis to quadriplegia and incontinence. Thus management strategies are very important to ensure 2 essential requirements: that the compression at the Cervico-Medullary Junction is released, and stability of the Cranio-Vertebral Junction is ensured.

We present a patient with improper correction of an AAD who presented with inadequate decompression with adequate stabilization, posteriorly with a Gallies fusion. The patient had no symptomatic relief despite aggressive physiotherapy. Imaging revealed good bony fusion but severe compression on the cord by a large jutting odontoid. The odontoid was removed trans orally and the patient was subjected to rehabilitation therapy. She improved well and is ambulant without support now.

The report highlights the need to address both aspects of cranio-vertebral junction pathology when confronted with a case as mentioned. Simply stabilizing the C1-C2 joint or C0-C2 joint without adequate neurological decompression is NOT enough. Tailoring the treatment to meet the needs of the patient is essential for proper patient recovery and satisfaction.

Keywords: Atlanto-axial dislocation, Odontoid, Quadriplegia, Instability

INTRODUCTION

Atlanto-Axial Dislocation is rare, yet devastating to its sufferers. Congenital AAD is even more infrequent, arising out of insufficient maturation and development of the branchial arches and occipital and cervical somites [1-4]. Other causes of AAD such as inflammatory diseases, trauma and bone disorders are more common and increasing in incidence. The primary problem with an AAD is the compression on the Cervico-Medullary Junction, critical to vegetative functions such as Respiration, Cardiac activity and consciousness [3-5]. Time and again, mortality and severe morbidity has been demonstrated due to dysfunction of the CMJ alone [3-5] Yet reams of literature have been published focusing on the dangers of instability at the CVJ. This has led to a disproportionate emphasis on fusion of the CVJ rather than decompression of the CMJ [5]. The result although an improvement from before, leads to little improvement and long standing damage to the medulla and cervical cord leading to sometimes irreversible damage to the neural elements of the CMJ. It is therefore critical to examine both aspects carefully before embarking on a surgical strategy for the patient. Such a case is discussed below.

CASE REPORT

A 62 years old lady was admitted with spastic Quadriplegia (2/5) and urinary urge incontinence. Her neurological status had worsened following a trivial fall 10 days ago. She had presented with the same problem 10 years ago when she was seen in 1994. She was diagnosed with congenital Atlanto-Axial Dislocation (AAD) and subjected to an Occipito-Cervical Fusion with a posterior stainless steel wiring and bone graft construct. Post-surgery her spasticity was the same although her neck pain was much better. She was subjected to rigorous physiotherapy but too significant improvement. She was, before her fall, ambulant with support and able to perform her activities of daily living with

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minimal assistance.

On arrival in the OPD, she was bedridden with spastic quadriplegia Power grade 2/5 with spasticity of 4(modified Ashworth's score). Her lower cranial nerves were normal allowing her to swallow and speak normally. She had no radicular pain, but complained of paraesthesia over all 4 limbs all the time.

On investigating her situation, her X-ray of the cervical spine and CVJ (**Figure 1**) showed good bony fusion with the stainless steel wire in situ posteriorly. Dynamic X-rays showed no instability, but Basilar Invagination was seen. A CT scan of the cervical spine and CVJ confirmed the findings of the X-ray. This showed Basilar Invagination (BI) and an increased Atlanto-Dens Interval (ADI) (**Figure 2**).

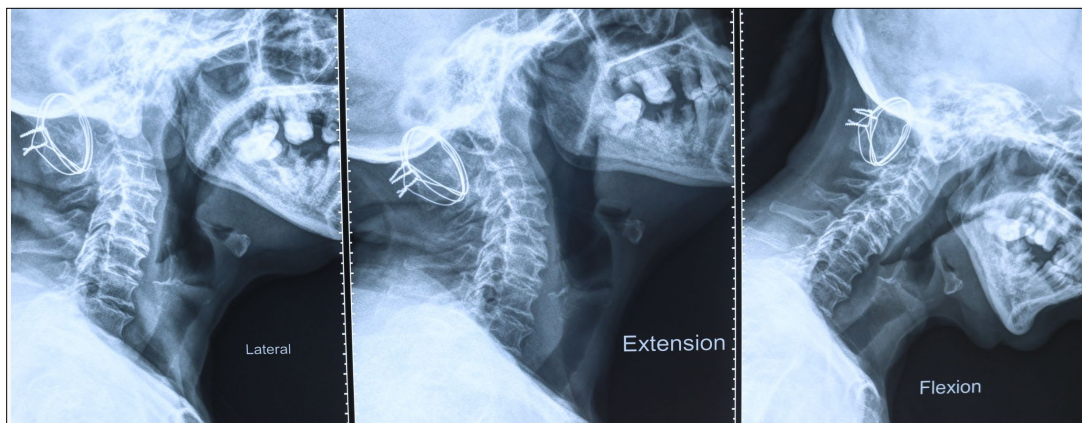


Figure 1. Dynamic X-ray of the cervical spine and CVJ showing good bony fusion between C0, C1 and C2 with steel wire *in situ*.

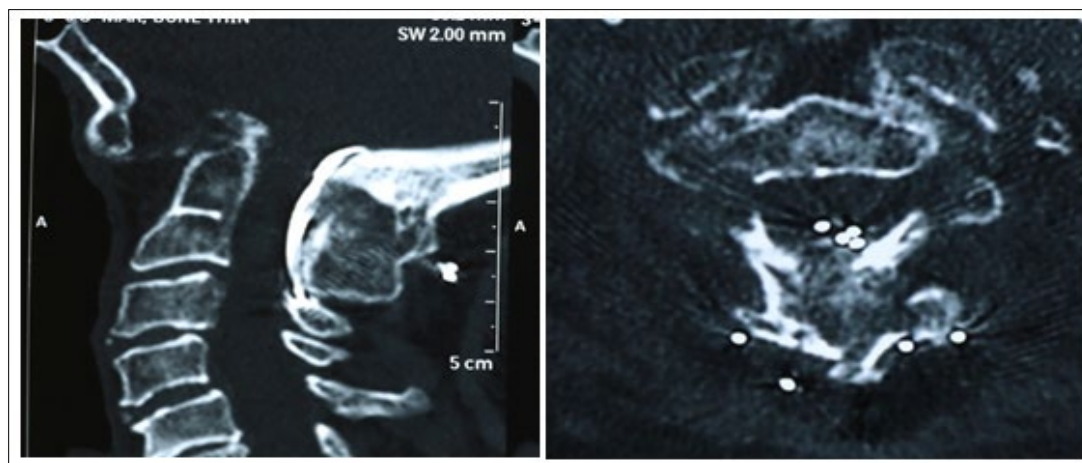


Figure 2. CT C-spine showing basilar invagination and increased atlanto dental interval (ADI) (a) with stainless steel wire *in situ* (b).

Hence it was clear that the fusion done 10 years ago did only convert a mobile AAD into a fixed AAD. There was no correction of the BI either which had led to a fusion of the C0-C1, C2 complex in a compromised position. The next part of our investigation was to probe the extent of damage sustained by the Cervico-Medullary Junction (CMJ) and Spinal Cord. This proved difficult, as the stainless steel wire prevented us from getting an MRI of the CVJ done. Due to the long-standing disease and wrong surgery done in the past, we concluded that the MRI was a priority, hence the stainless steel wire was surgically removed (as fusion had occurred, its function *in situ* was void) and the patient was then subjected to an MRI of the CMJ and Cervical spine.

The MRI showed massive cord compression at the CVJ with thinning of the CMJ and myelomalacia changes. No Chiari, hydrocephalus, syrinx or other abnormalities were seen (**Figure 3**). The dire nature of the compression, primarily due to the AAD and the BI which had not been managed appropriately before, placed us in a dilemma regarding the high risk, the decompression now entailed. Resultant cord edema, could lead to catastrophic respiratory paralysis and prolonged ICU and Ventilator dependency, not to mention quadriplegia, and death. To be doubly sure of the stability of the CVJ, another dynamic X-ray was taken which showed good stability (**Figure 4**).

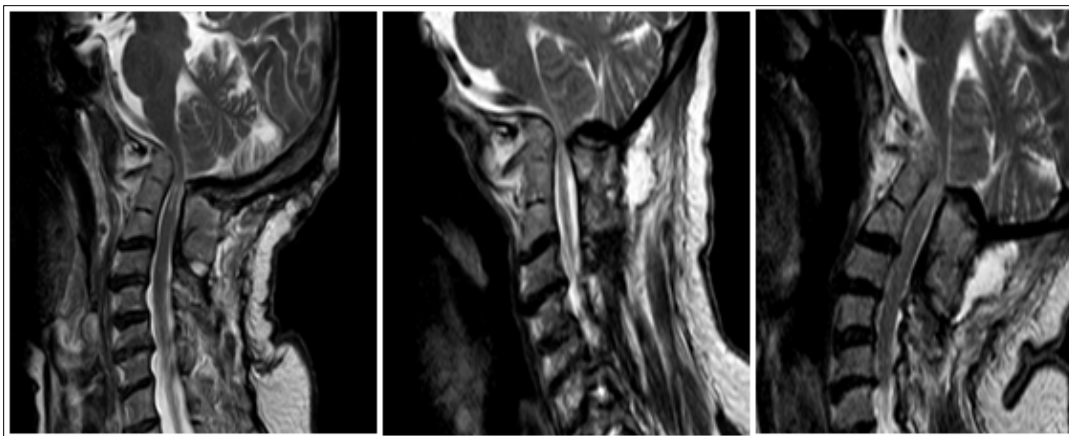


Figure 3. MRI C-spine in flexion and extension showing untreated severe CMJ compression with myelomalacia.

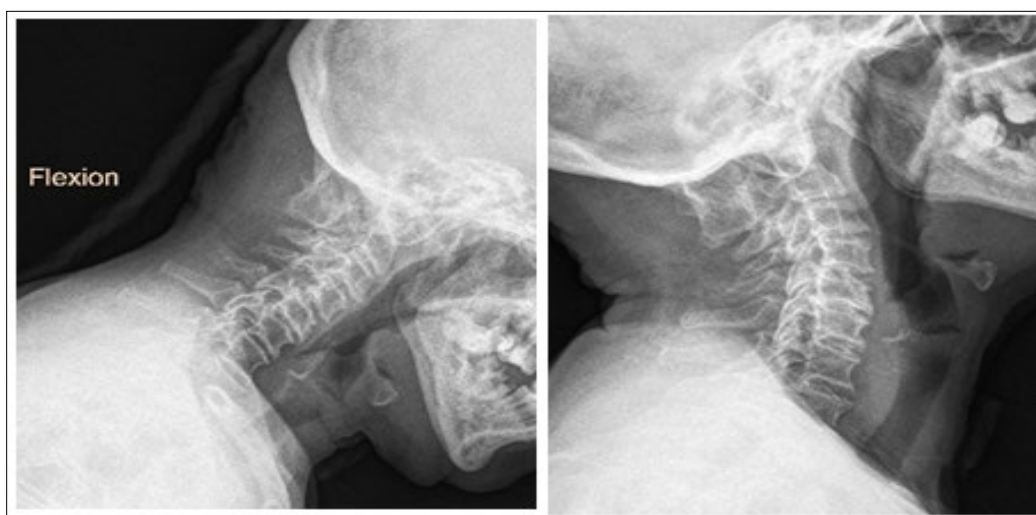


Figure 4: Flexion extension x-ray of the C-spine after removal of stainless-steel wire showing good bony fusion.

The options before us were:

1. Foramen Magnum Decompression (FMD)
2. Trans-Oral Odontoidectomy

We proceeded with the latter option, as the primary cause of compression was the malpositioned odontoid process of C2.

After a thorough work up, the patient was placed supine after nasopharyngeal intubation was done and mouth opened with a Boyle-Davis Mouth gag. The posterior pharyngeal wall was incised and dissected under C-ram guidance to reveal the anterior arch of C1 and the Odontoid. Both were drilled to expose the dura of the CMJ. Haemostasis was achieved, before closure was done with soluble sutures.

Post op the patient was mobilized immediately and she showed significant decrease in spasticity. Gradual physiotherapy and rehab care showed excellent recovery. She now is ambulant without support, and performs all activities of daily living freely. She is also active in the

kitchen and performs her daily chores well (something that was impossible pre-op) and lives a happy functional life.

DISCUSSION

Craniovertebral Instability is a difficult problem to address. No two patients are the same. Tailoring the management to each individual's needs is important to ensure good recovery and patient satisfaction [5-7]. The most important aspects of CVJ disorders are that, there are 2 main issues that need attention.

Instability [1-11]

The improperly fused elements of the CVJ lead to instability in weight bearing of the head. Such a problem is significant and alarming, which can lead to significant morbidity. However, CVJ instability is not seen in all disorders. Its severity and the need to intervene, needs to be assessed with respect to the pathology of the disease and the cause of the symptoms of the patient. Blindly fusing all CVJ abnormalities will result in patients like the one discussed,

where fusion and excellent stability have resulted in no improvement in functional status.

Neurological decompression [4-6,8,11]

The crux of all procedures, aimed at resolving disease at the CVJ, eventually involves reduction of compression (either bony, ligamentous or produced by external agents such as pannus or pus) on the CMJ. Without adequate neurological decompression, there will be no symptomatic relief for the patient and hence no recovery. It is therefore imperative to understand the nature of the disease and the cause of the symptoms of the patient.

Deciding upon the right management strategy requires many stages of deduction to be performed. These include [7-12]:

1. **Neurological history:** These help deduce the causative pathology and thereby point in the direction of the plausible problem. For example, traumatic CVJ dislocations will usually have a predominant component of instability rather than compression while rheumatoid arthritis and inflammatory conditions, will lead to pannus formation and predominantly compressive pathology. A good grasp of the causative etiology and the pathological progression helps decide on surgery, and adjuvant therapy [10-12].
2. **Examination:** The presence of a Myelopathy definitely implies serious neurological compression, making its release a priority [12-14].
3. **Presence of abnormal anatomy:** The existence of a BI, AAD or other related abnormalities implies reduction to normal anatomical position along with adequate decompression [6,8,11,12].
4. **Instability and neurological compression seen on neuro-radiology:** The need for a dynamic study of the CVJ is obvious as it detects instability best of all. In certain complicated anatomical situations, CT scan in flexion and extension is helpful to identify the chief offending pathology. An MRI however is essential to detect the status of the CMJ and the cervical cord. No manipulation of the CVJ and CMJ must be done without an MRI. Failure to do this can result in catastrophic destruction of a delicate and vital structure [3,5,8,9,11,13,14].
5. **Presence of previous surgical implants and their implications to management:** As discussed in the case report, the presence of the inter laminar fusion wire along with posterior fusion changed the situation completely. Although trans-oral odontoidectomy is frowned upon nowadays, due to a sturdy posterior fusion being present, there was no need to re-stabilize the joint. Hence only decompression was required. The stainless-steel wire caused us much grief, as an MRI of the CMJ was impossible with it in situ. Due to the essential nature of the MRI, it was decided to surgically

remove it and then take an MRI before planning definitive management. Such a strategy, although cumbersome and long-winded, will eventually point the correct direction to take, and hence is worth the trouble to both patient and surgeon.

The algorithm for management is well known and is hence not addressed here. Posterior fusion with adequate decompression is the treatment of choice here. The choice of C0-C1-C2 fusion or just C1-C2 fusion depends upon the extent and nature of the instability. Decompressive procedures include laminectomies and Foramen magnum decompressions. These again are to be decided upon studying the MRI and matching the compression to the symptoms experienced by the patients as cause and effect.

Lastly, postoperative physiotherapy is perhaps more important than surgery itself [14]. Rehabilitation experts are the wizards who make noteworthy improvements in the lives of the patients. Surgery alone is useless if not combined with proper rehab support. As with our patient, it was long-term, persistent and assiduous rehab care that led to the excellent results experienced by our patients [14]. This underlines the need of a team approach to these complex conditions, where Neurosurgeons, nurses, physiotherapists, anesthetists, radiologists and technicians combine to give the patient the best result possible.

SUMMARY AND CONCLUSION

Pathologies of the CVJ are complex and do NOT have a 1 size fits all solution. A thorough understanding of the situation coupled with adequate emphasis to BOTH neurological decompression AS WELL AS instability gives the best result overall, and help avoid difficult and inconvenient redo surgeries.

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