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Case Report

Occipitocervical Fusion for Atlantoaxial Instability Secondary to Post Laryngeal Procedure Infection in a Down Syndrome Patient - Case Report

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Abstract

Atlantoaxial instability (AAI) and subluxation may be associated with a variety of diseases. These include congenital and acquired conditions. The latter includes infectious or inflammatory processes, especially coming from the retropharyngeal space (Grisel syndrome, GS), which frequently affects children. This population presents with anatomical features and an increased frequency of upper respiratory infections, which explains the higher incidence between them. The risk for C1-C2 subluxation can be further increased by the presence of congenital ligament laxity, such as in individuals with Down syndrome. GS treatment often involves antibiotic therapy (in the setting of an acute infection) and close subluxation reduction and cervical stabilization. Surgery is reserved for highly unstable spine or failure after non-operative management. Here, we present a case of a 7-year-old child with destructive infectious AAI after a laryngeal procedure to treat subglottic stenosis. The concomitant presence of Down Syndrome (a congenital cause of AAI) led to a highly unstable spine, with neurological deficits and destruction of C1 lateral mass, that required upfront open reduction and fixation to warrant function preservation. Given the uncooperative and agitated patient, conservative management was deemed risky and would not prevent further subluxations in the future. The patient had an uneventful recovery period and remained neurologically stable at her last outpatient visit, with no signs of active infection. We concluded that the decision-making process for conservative versus surgical management for infection-related atlantoaxial instability is complex and must consider the individual characteristics and also neurological risk.

Keywords: Grisel; Syndrome; Spine; Down; Fusion; Atlantoaxial

Abbreviations

AAI: Atlantoaxial Instability; DS: Down Syndrome; GS: Grisel Syndrome; MRI: Magnetic Resonance Imaging

Introduction

Atlantoaxial instability (AAI) may relate to a variety of conditions, including genetic, rheumatologic, and infectious diseases [1]. It also affects 10% to 20% of all individuals with Down Syndrome due to a high risk of congenital increased mobility between the C1 and C2 vertebrae [2]. In these cases, although relatively common, it is mainly asymptomatic and often diagnosed through radiographs ordered for another non-related condition [2].

The bone and ligament destruction caused by local infections and inflammatory response also leads to C1-C2 ligaments and joint destruction and consequent instability [3,4]. Infection may come from the contiguous spread of pathogens from upper respiratory infection or from a head and neck surgery complication [4]. In general, it is polymicrobial and presents with cervical pain, restricted neck mobility, fever, cervical lymphadenopathy, odynophagia, and dysphagia. The early infection recognition and prompt treatment are of great importance in avoiding catastrophic complications [1,4].

In this paper, we present the case of a child with multi-factorial AAI (ligament laxity, due to Down Syndrome, and destructive osteomyelitis, from a contiguous infection after a laryngeal procedure) and its treatment. Consent was obtained from the parents.

Case Report

A 7-year-old girl, previously diagnosed with Down Syndrome (DS) with associated moderate cognitive impairment, was brought to the emergency room with a 30-day history of opioid-refractory neck pain with painful restriction to head rotation and fixed head position. Additionally, she presented hypoactivity, eating avoidance, and fluctuant fever. At initial neurological evaluation, there was tetra paresis (grade III in both superior and inferior limbs) and mild spasticity. Past medical history included recurrent pneumonia, and a laryngotracheoplasty performed three months before current symptoms appeared, for the treatment of a subglottic stenosis – she was using a tracheostomy tracheal tube.

Radiographs and magnetic resonance imaging (MRI) were ordered (Figure 1). These have shown vertebral osteomyelitis with destruction of the right C1-C2 facet articulation with mobile anterior displacement of C1 over C2 and cord compression. There was a retropharyngeal abscess communicating with the spine.

These findings warranted prompt sedation and mechanical ventilation given the imminent risk of progressive deficit in this agitated and uncollaborative patient. A Philadelphia cervical collar was placed.

Despite the local infectious process, we felt that the patient would benefit from urgent occipitocervical fusion to block the progression of the ongoing spinal cord compression. Under general anesthesia, the patient was positioned prone with a slight head extension, which was fixed with a three-pin Mayfield headholder. With this, the partial correction of the subluxation was obtained, and this was confirmed with fluoroscopy (Figure 2). A posterior midline incision was made over the inium to the C4 vertebra level. With subperiosteal dissection, the paravertebral muscles were retracted, and the occipitocervical bony elements exposed (Figure 3). The dissection of the soft tissue at the right side of the C1-C2 levels led to a purulent content, which was drained and cultured (resulted in no microorganism growth). A titanium plate was fixed to the occipital bone and titanium screws were placed on the left C2 pars articularis and right C2 lamina. The posterior C1 arch was removed, and the system was connected through titanium rods and a cross-linking piece (Figure 4).

After the spinal procedure, the patient was flipped over and a

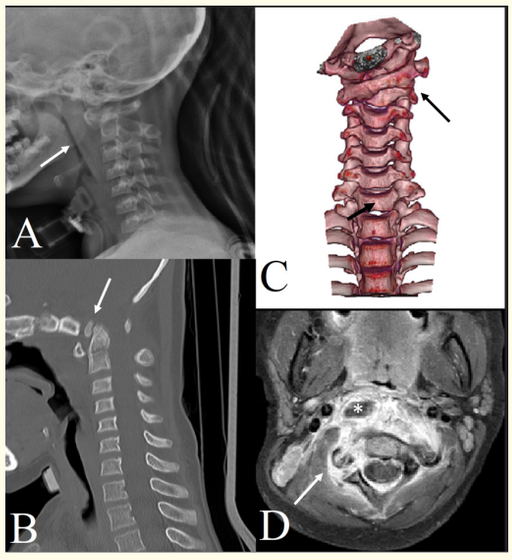

Figure 1: A, cervical lateral radiograph. The atlas (C1) is anteriorly displaced over the axis (C2), with an increased atlas-dens interval, and there is an increased diameter of the prevertebral soft tissues (white arrow). B, computed tomography confirming radiograph's findings. Additional finding of os odontoideum (white arrow). C, computed tomography 3D reconstruction showing the subluxation affecting mainly the right side of C1- as the left articulation is relatively preserved (black arrow), this served as the pivot for the translational movement of C1. D, T1-weighted MRI showing contrast enhancement in the prevertebral soft tissues, extending to the right portion of C2 (white arrow). A pus-containing cavity can be seen on the right (asterisk).

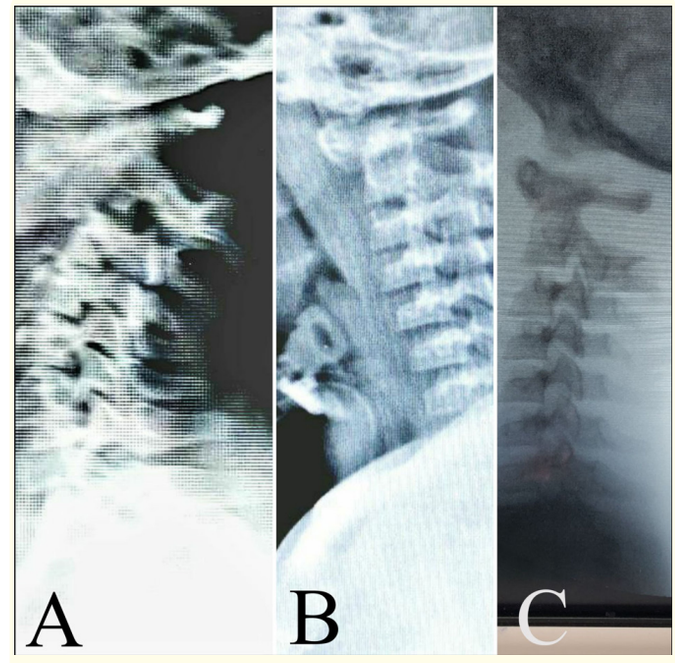

Figure 2: A, fluoroscopy, lateral view. Subluxation of C1-C2 before closed reduction. B, fluoroscopy, lateral view after a cervical collar and sedation. Almost all the subluxation was reduced with proper manual head position.

Figure 3: A, planned skin incision. B, the head was positioned slightly extended and fixed with a three-pin Mayfield headholder. C, intraoperative fluoroscopy showing reduction of subluxation. D, intraoperative photograph depicting the exposure of the occipitocervical region's bony elements.

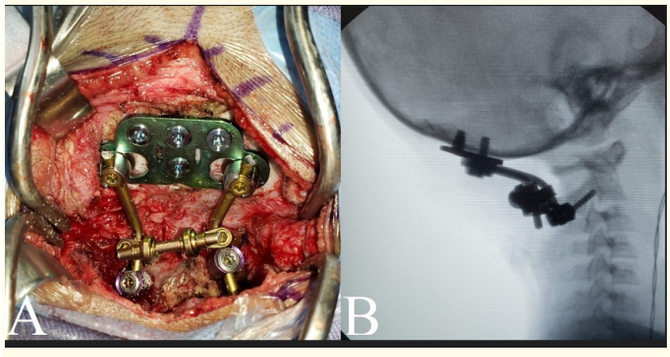

Figure 4: A, intraoperative photograph showing the final product. B, intraoperative fluoroscopy, lateral view, demonstrating the resolution of the atlantoaxial subluxation and the titanium hardware.

transoral abscess puncture was performed, and additional purulent material was collected. Cultures demonstrated methicillin-sensitive *Staphylococcus aureus* and *Candida albicans*, which were treated with intravenous clindamycin and fluconazole.

In the postoperative period, sedation and mechanical ventilation were weaned off. She presented a significant improvement in pain and a preserved neurological function. The C-reactive protein levels had a progressive and constant decline during her hospitalization. She was discharged 4 weeks after surgery and antimicrobial therapy with oral sulfamethoxazole-trimethoprim (planned for a six-month period).

At the first outpatient clinic visit, the patient was pain-free, with a mild residual spasticity, and no signs of hardware-related infection. She was followed up for more than six months with no recurrence of infection and no neck pain, able to freely walk without assistance.

Discussion

A variety of conditions including congenital, infectious, and rheumatologic diseases may lead to AAI [1]. Nontraumatic atlantoaxial rotatory subluxation has been associated with head and neck infections and surgery, the also called Grisel syndrome (GS) [4]. Previous studies on the craniocervical junction anatomy have shown the presence of a periodontoidal vascular plexus draining the upper posterior pharyngeal region, which promotes the easier propagation of retropharyngeal septic exudates to the spine [5]. GS affects mostly children, with around 70% occurring in individuals younger than 12 years [1,3,6]. The classical clinical presentation is a painful torticollis with or without fever, and may be associated with neurological findings such as those related to spinal cord compression or cranial nerve palsy [7]. Initial workup includes plain radiograms or a CT scan (gold standard) of the craniocervical junction [7]. By the 3D reconstruction of CT images, it is possible to classify the disease according to the Fielding-Hawkins classification [8]. Our patient had a subluxation that was classified as type II, given its intact left C1-C2 facet articulation that served as the pivot for the translational movement. MRI provides important information on the soft tissues, lymphnodes, spinal cord compression status, and infectious process extension [7].

Similar to other studies' findings, *Staphylococcus aureus* has been isolated in our case's abscess aspirate [1,6,9]. The presence of *Candida* was probably contaminant [10], but warranted treatment given the disease's severity. It was initially thought that the infectious/inflammatory process would lead to a ligament softening through hyperemia, however, other authors suggested that the muscle spasm secondary to infection in association with a previous ligament laxity (i.e., two-hit hypothesis) is probably the real cause [11]. Children would be more vulnerable to GS due to their relative weaker ligaments, higher frequency of upper respiratory infections, immature bone formation, occipito-atlanto-axial joints with larger synovial folds, adeno-tonsillar hypertrophy, horizontally-oriented facets, and insufficient cervical muscle strength [3,4,7]. In addition to that, our case had DS, which is a known cause of ligament laxity and may present with spontaneous atlantoaxial subluxation [2,7]. The combined presence of these factors led to a highly unstable

spine, with associated spinal cord compression.

The standards for its management are still not clear [7]. Based on reported successful results, some authors have suggested that this condition may receive initial conservative management, with antibiotics and external reduction and immobilization [1,3,12]. In more severe instability cases, the use of a halo may be considered [5]. Around 12% of children with GS will require surgical stabilization, either as first- or second-line treatments, and is usually recommended in cases refractory to conservative management or highly unstable spine cases (Fielding-Hawkins grade IV) [1]. This is usually obtained by the placement of metallic implants attached to the spine by screws, whose hardware construct length and configuration may vary. We opted for an occipital-C2 fusion skipping C1 given the lateral masses involvement in the infection process. Even though there is a fixation-related head rotation range of motion reduction, open surgery provides an early recovery and prevents recurrence [7]. This rigid spinal stabilization is of special interest in patients with an increased ligament laxity baseline, such as in DS. For this reason, in combination with the patient's agitation, we felt that open spinal reduction and fixation would warrant long-term stability while protecting against further neurological deterioration, as well as allowing immediate drainage of the pus collection. To the best of our knowledge, this is the second case presenting a combination between GS and DS in children in the literature [13], and the first one to demonstrate the successful use of an occipito-C2 fusion to treat Grisel syndrome in the same population.

Therefore, the most adequate management to GS must take into consideration multiple factors, including individual's comorbidities and neurological status. Despite evidence favoring the use of a stepwise approach, in a gradual invasiveness increase [7], for most of the cases, some patients may benefit from an upfront surgical stabilization.

Conclusion

We presented the case of a 7-year-old child with atlantoaxial subluxation related to a combination of risk factors (DS and retropharyngeal infection). The upfront surgical occipital-cervical (C2) fusion prevented neurological function decline and offered long-term stabilization to this high-risk patient. This is the first paper to report its successful application in this population. Our results suggest that the decision-making process for conservative versus surgical management is complex and must consider the individual characteristics and neurological risk.

Conflict of Interest

Authors disclose no conflict of interest.

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