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Case Report

Recurrent Torticollis and Cervical Subluxation in a Pediatric Patient

Hamail Iqbal[1] and Catherine A. Mazzola[2*]

[1]Rutgers University, New Brunswick, New Jersey, USA
[2]New Jersey Pediatric Neuroscience Institute, USA

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ABSTRACT

Background: Atlantoaxial subluxation (AAS), also referred to as C1-C2 subluxation, is a misalignment of the first two vertebrae of the cervical spine. AAS typically presents with a head tilt (torticollis) with limited range of motion (ROM). Torticollis is quite common in infants, but in older children, torticollis may be an indication of AAS.

Method: In this retrospective case study, the clinical history of a female pediatric patient diagnosed with atlantoaxial subluxation presenting with recurrent torticollis is reviewed.

Result: The patient was initially diagnosed with torticollis during infancy; torticollis partially resolved. However, after an ear infection, the child again developed torticollis secondary to Grisel's Syndrome. Despite undergoing physical therapy treatments, torticollis persisted. The patient was referred to neurosurgery at age nine. AAS was diagnosed after a three-dimensional (3D) computed tomography (CT) scan. The child was subsequently placed in halo-traction to reduce the C1-C2 subluxation. Once the alignment was acceptable, the child was placed in a halo vest. However, even after several months of non-invasive cervical spine immobilization with a halo vest and hard cervical collar, the head tilt and cervical subluxation recurred due to bone remodeling.

Conclusion: The diagnosis of AAS requires both a comprehensive physical examination and imaging following presentation of torticollis. Understanding the etiology of the torticollis early on is critical in preventing the occurrence of AAS after treatment.

Introduction

Atlantoaxial subluxation (AAS), also known as C1-C2 subluxation, is a dislocation of the first cervical vertebra over the second cervical vertebra. AAS impairs the full range of motion (ROM) of the neck. Acute cervical spine trauma is the most common cause of C1-C2 subluxation; however, non-traumatic causes have also been widely reported [1]. Specifically, Grisel's Syndrome (GS) is a rare condition marked by inflammation of neck tissue surrounding the atlantoaxial joint secondary to infections of the head, neck, or upper respiratory tract [2]. One of the most common findings of C1-C2 subluxation in physical examination is torticollis, an abnormal tilting of the head to one side. Although common in infants, in an older child, torticollis can indicate cervical spine instability or underlying muscle or bone (vertebral) deformities as well [3]. Due to the variability in torticollis etiologies, early diagnosis and treatment are essential in preventing progression of instability, subluxation, and optimizing the effectiveness of non-surgical and surgical treatment of C1-C2 subluxation.

Case Report

At 6 months of age, a female patient who was born full term without complication, was diagnosed with torticollis. Shortly after, she was diagnosed with torticollis secondary to an ear infection. No further testing or treatment was recommended. At 2 years of age, the patient was seen by a neurologist for persistent torticollis, and a magnetic resonance imaging (MRI) study of the brain was recommended. The MRI was "unremarkable", and torticollis was felt to be musculoskeletal. The patient was referred for physical therapy and prescribed a tubular

*Correspondence to: Catherine A. Mazzola, M.D., FAANS, New Jersey Pediatric Neuroscience Institute, USA; E-mail: cmazzola@njpni.com

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orthosis for torticollis (TOT) collar to be worn three times a week. Cervical spine radiographs were also performed around this time; however, the results of these images were felt to be "normal" and no further treatment was recommended.

After 2 years of physical therapy, at age 4 years, there was no significant improvement in the patient's torticollis and the treatment was stopped. Subsequent medical history revealed that the patient underwent a tonsillectomy at 6 years of age and presented to a pediatrics office visit one year later, in July 2017, again with severe head tilt and right-sided neck pain. According to the patient, the pain began one year earlier, concurrent with prominence of lymph nodes in the same region. The physical examination performed at this time determined torticollis secondary to a tight right sternocleidomastoid (SCM) muscle and was consequently referred to pediatric neurosurgery to discuss options for botulinum toxin injections or surgical intervention, at age nine.

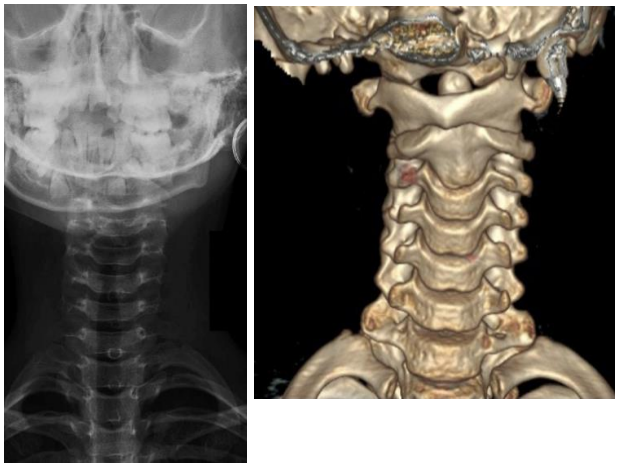

Figure 1: A) Anterior radiograph of the cervical spine performed in 2018 demonstrating normal alignment of visualized cervical spine, but C1 and C2 are not well seen. **B)** Anterior view of three-dimensional reconstruction of CT scan done in 2019 demonstrating C1-C2 subluxation.

On examination, the patient was a well-developed and normal appearing nine-year-old girl, who had an obvious fixed torticollis. On presentation, she had limited range of motion, but could initiate movement and rotation to the right and left, with a preference to tilt the head towards the right. Intermittent paresthesia and numbness of the extremities and episodes of loss of balance were also reported at this time. Radiographs of the cervical spine, done in early 2018, did not detect any acute fracture or subluxation (Figure 1). However, the C1-C2 vertebrae were not adequately visualized. Then a cervical spine MRI performed four months later raised suspicion for potential C1-C2 subluxation. A follow up computerized tomography (CT) scan with three-dimensional (3D) reconstruction performed one month later clearly documented rotation of C1 relative to C2 as well as bone remodeling of the C1-C2 joint (Figure 1). Consequently, a halo with eight pin fixations (with 6 pounds per square inch of pin torque) was placed. The child was placed in halo traction for several days. Once C1-C2 subluxation resolved and normal alignment was obtained, the child was placed into a halo-vest, a cervical spine immobilization treatment. The halo-vest was maintained for approximately two and a half months. Radiographs taken intermittently over the duration of halo-vest use demonstrated normal alignment of C1 on C2 with decreasing odontoid and lateral mass asymmetry (Figure 2).

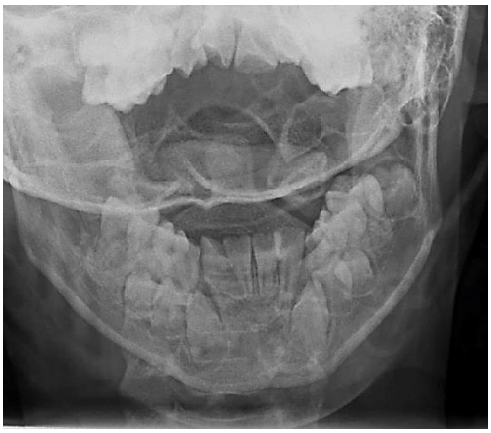

Figure 2: Open-mouth radiograph of cervical spine, performed during halo-vest immobilization, demonstrating normal C1-C2 alignment.

Following removal of the halo-vest, the patient was placed into a Miami J hard collar for two months and physical therapy was started. After discontinuation of the hard collar, placement of a soft cervical collar, and continuation of physical therapy, the patient began to develop recurrent torticollis. Significant head tilt returned about one month after removal of the hard collar. The soft collar, worn by the patient after discontinuation of the hard Miami J collar, was found to be inadequate in ensuring proper head positioning. At that time, the patient's parents opted not to pursue surgical fusion of the C1-C2 vertebrae.

Discussion

This report describes the case of a female pediatric patient with recurrent torticollis present for several years. Though the initial diagnosis for torticollis was made at the age of six months, the patient still presented with a head tilt towards the right at the age of nine. Although her C1-C2 subluxation was reduced and corrected, proper alignment could not be maintained despite 2 months in a halo-vest and 2 months in a hard collar.

I Etiology

The child initially presented as an infant with torticollis. As a toddler, she presented again with an abnormal head tilt following an ear infection. At that time, her torticollis was likely secondary to GS. Battiata *et al*. describe drainage from the nasopharynx through the pharyngo-vertebral veins and to the peri-odontoid venous plexuses as a pathway for infectious or inflammatory contents to spread to the atlantoaxial ligaments [4]. As the Eustachian tube provides a direct pathway between the outer ear and the nasopharynx, it is reasonable to conclude that inflammation from the ear infection caused ligamentous laxity and, consequently, torticollis [5]. GS also presents after maxillofacial or otorhinolaryngology procedures, including tonsillectomy and adenoidectomy [2]. For the patient presented in this report, though the tonsillectomy was documented as having no postoperative complications, the onset of neck pain, as well as the finding of enlarged lymph nodes, around the same time as the tonsillectomy suggest recurrence or exacerbation of existing torticollis secondary to GS [6].

Another significant finding is the tightness of the right sternocleidomastoid muscle (SCM). The SCM allows ipsilateral-lateral flexion and contralateral rotation; accordingly, tightness of the right SCM caused pain when the patient turned her head to the left [7]. The patient's asymmetrical head positioning likely caused fibrosis and hypertonia of the right SCM due to inactivity [3, 7]. Missaghi describes the case of a 37-year-old female with gradually worsening unilateral neck pain after carrying her child for long periods of time with her left arm while performing tasks with her right arm; various physical examinations, including cervical flexion-extension and muscle strength and length tests, were performed leading to the determination of SCM tightness [7].

Furthermore, a few studies have described cervical spine vertebral and bone deformities secondary to muscular torticollis. Hussein *et al*. presents a case review of fifteen pediatric patients, all under the age of eight years, with congenital muscular torticollis (CMT), a condition caused by pathological shortening of the SCM. While the patients under one year of age did not show significant gross changes in cervical spine anatomy, older patients showed progressive tilting of the odontoid process and minimal rotation of the axis [8]. Thus, the C1-C2 subluxation found on the CT scan of the patient in this study may have been caused by chronic SCM dysfunction. It is important to consider, however, that the review of Hussein *et al*. and other similar studies reported specifically on CMT; the pathophysiology of cervical spine dysfunctions secondary to acquired muscular torticollis may be similar to that from CMT and requires further study.

II Radiological Methods

While an MRI cervical spine study introduced the initial suspicion of C1-C2 subluxation, the final determination was found only through CT. Though the high radiation exposure of CT scans presents a concern in the pediatric patients and their families, the high bone resolution is optimal for imaging patients at a high-risk of C1-C2 subluxation [9].

III Treatment

Over the course of her medical history, the patient in this report received multiple different therapies for torticollis and C1-C2 subluxation, such as physical therapy, TOT collar, halo traction, and halo-vest stabilization, followed by hard and soft collar stabilization. While initial physical therapy visits and TOT collar use failed to produce significant results, halo traction and halo-vest immobilization provided near-complete resolution of C1-C2 subluxation and torticollis. The gradual recurrence of the torticollis was likely due to insufficient collar support following halo-vest and hard collar removal. Another possible causation of recurrent torticollis is C1 and C2 joint and bone remodeling after long term subluxation. Since the inflammatory process of GS leads to a hyperemic state in the prevertebral tissue, progressive decalcification of C1 and C2 with joint remodeling occurs [4].

Conclusion

Atlantoaxial subluxation, from both traumatic and nontraumatic etiologies, presents serious consequences if it is not detected and treated promptly. As torticollis is one of the main clinical findings of the condition, physical evaluation and radiological studies should be performed at the first presentation of the symptom to rule out C1-C2 subluxation. The most effective treatment for torticollis secondary to C1-C2 subluxation is cervical spine reduction and immobilization in a hard collar or halo-vest. However, the presence of other musculoskeletal risk factors may require prolonged immobilization and monitoring of cervical alignment.

Abbreviations

AAS: Atlantoaxial Subluxation
CMT: Congenital Muscular Torticollis
CT: Computerized Tomography
GS: Grisel's Syndrome
MRI: Magnetic Resonance Imaging
ROM: Range of Motion
SCM: Sternocleidomastoid
3D: Three Dimensional
TOT: Tubular Orthosis for Torticollis

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