A 17-year-old girl presented to the emergency department with torticollis toward the left. Three months prior to presentation she had suffered from tonsillitis and awoke one day with torticollis, denying any history of trauma. Initial symptoms included nonspecific sensory symptoms involving the upper limb and pain in the neck with restriction of neck movements. The tonsillitis settled uneventfully with antibiotics. She consulted her general practitioner who managed her torticollis conservatively with physiotherapy and manipulation with no relief.

On examination, there was fixed rotation of the neck to the left with normal neurological assessment. Active manipulation of the head and neck was painful and therefore reduction impossible. A computed tomography (CT) scan and magnetic resonance imaging demonstrated rotatory subluxation at C1/C2 with intact ligaments and no critical compression of the spinal cord [Figures 1 and 2].

An attempt at manual reduction was unsuccessful, so Gardner-Wells tongs with traction weight was applied with regular oral diazepam concurrently. Four days later and with 8 pounds (3.63 kg) of traction weight, a CT scan and plain radiographs of her neck confirmed near normal alignment [Figure 3].

She was, therefore, braced with a pinned HALO vest and followed-up closely as an outpatient. After a total of 3 months.
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in the pinned HALO, she was transitioned into a “cyber” neck support and subsequently placed into an aspen collar which was also gradually weaned off over a period of 4 weeks. She remains neurologically intact with normal radiological vertebral alignment.

Discuss

The atlantoaxial joint is formed by two opposing, saddle shaped biconvex surfaces which allows up to 40° of rotation to either side. The most widely accepted theory for Grisel’s syndrome is the two-hit hypothesis. The first-hit is cervical ligamentous laxity seen in the pediatric population at baseline; the second-hit is the induction of spasm by inflammatory mediators carried to cervical muscles by the pharyngovebral venous plexus.

Fielding and Hawkins described a grading system with rotatory fixation that serves as a guide to the severity of injury:

- Type 1 is <3 mm anterior displacement of the atlas
- Type 2 is 3–5 mm displacement
- Type 3 is >5 mm and
- Type 4 is posterior displacement.

Types 1 and 2 are not typically associated with neurologic impairment, but Types 3 and 4 are reported to have a 15% incidence of a range of neurologic involvement, the most severe being paralysis and death.

Kraft and Tschopp have proposed to add Type 0 - persistent torticollis with no radiologic evidence of subluxation.

Treatment suggestions are

- Type 0 - antibiotic therapy with close follow-up
- Type 1 - conservative with bed rest, oral nonsteroidal anti-inflammatory drugs, muscle relaxants, reduction (if required) and immobilization with a soft collar
- Type 2 - reduction and cervical traction with a stiff cervical collar followed by a period of the halo vest for 6–12 weeks
- Types 3 and 4 - cervical traction and bed rest with a period of cervical immobilization.

All the above would require serial imaging in order to assess progress. Surgery with open reduction and cervical fusion should be explored in recurrent cases or where conservative treatment has failed. The patient discussed here had a Type 1 injury, but due to the long interval between onset and presentation, a period of traction and HALO were applied. This case illustrates that there can be a variety of presentations of atlantoaxial rotatory subluxation, and each case should be assessed carefully to determine the best management option for optimal patient outcome.

References


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