Case report

Grisel's syndrome associated with tonsillitis

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A B S T R A C T

Non-traumatic subluxation of atlanto-axial joint known as Grisel's syndrome is a rare condition. The pathogenesis of Grisel's syndrome is not clear but it seems laxity of cervical ligaments in children and an inflammatory process in neck might cause the syndrome. Here we present a case of Grisel's syndrome, a 9-year-old boy presented with torticollis and discuss about clinical radiological and treatment aspects of the syndrome.

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Introduction

Grisel's syndrome is a non-traumatic subluxation of the atlanto-axial joint with inflammation in adjacent soft tissues. Pathogenesis of syndrome is incompletely understood but an inflammatory process in neck can be the cause of Grisel's syndrome [9]. This syndrome is not triggered by trauma and patients have a history of head, neck infections or otorhinolaryngology procedures. It often affects pediatrics and 90% of patients are under 21 years old [1]. Here we present a case of atlanto-axial subluxation involving torticollis.

Presentation of case

A 9-year-old boy presented to emergency. He had no history of trauma, sore throat, otitis, dysphagia, drooling, fever and another systemic symptom.

Physical examination revealed no issues in neurological function such as assessment of muscle power, cranial nerve functions, tendon reflexes, and sensory deficit. No cervical spine tenderness was noted. He had limitation in cervical range of motion including rotation, flexion and extension and tenderness of soft tissue on left side of his neck without signs of inflammation.

Physical examination of pharynx had no signs of uvula deviation, asymmetric tonsils and bulging of posterior pharyngeal wall.

Paraclinical investigations demonstrated leukocytosis (11,000 per microliter), neutrophilia (60%), elevated ESR (124 mm/hr) and CRP (44 mg/L). A lateral neck x-ray for assessment of prevertebral soft tissue was done that was normal. Computed tomography (CT) scan of neck and brain with and without contrast revealed left sided mastoiditis (Fig. 1). Per clinical assessment, he had no evidence of mastoiditis. Patient was apparently undertreated with oral antibiotics as an outpatient.

We repeated brain and neck CT scan with and without contrast. Because of continuance of torticollis, Mastoiditis, posterior neck angle lymphadenopathy, adenoid hypertrophy and prevertebral soft tissue cellulitis in anterior portion of second and third cervical vertebral bodies without involvement of parapharyngeal space reported. Antibiotic therapy with Clindamycin and Ceftriaxone was applied but after sixteen days despite the experimental response no clinical response was founded. According to neurosurgical counseling we did temporal bone MRI with/without gadolinium injection (Fig. 2). Mastoiditis, osteomyelitis of first cervical vertebra, soft tissue cellulitis of prevertebral portion of first cervical and mild change in left petrous apex reported. To assess additional probable problem and need for fixation of neck we did neck CT scan with and without contrast with reconstruction of sagittal, coronal plane and three dimensions (Fig. 3). First and second cervical vertebral subluxation was diagnosed. The patient was treated with halter neck traction for three days and subluxation improved and treatment with orthosis continued for 3–5 months. For treatment of osteomyelitis we gave Ampicillin-sulbactam for one month. With experimental and clinical recovery, the patient with recommendation to use orthosis for 3–5 months and oral AB with Clindamycin and Co-amoxiclav for 4 months discharged.

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Discussion

Grisel’s syndrome is a rare condition usually affects children. Patients with Grisel’s syndrome typically present with torticollis in 15% of cases. It can be accompanied with neurological symptoms. Neurological symptoms range from upper limb numbness to bladder dysfunction, quadriplegia and respiratory failure. Etiology and pathogenesis of Grisel’s syndrome is unclear but it has been demonstrated than an inflammatory process in neck soft tissues like peritonsillar, retropharyngeal abscess or post-operative inflammation after adenoidectomy can lead to laxity [5]. Pharyngo-vertebral veins that drain nasopharynx are directly connected with periodontoidal venous plexuses. So, any inflammatory process in pharynx can transport to atlanto-axial ligaments. Any syndrome that involve ligaments such as Down’s syndrome can lead to Grisel’s syndrome. Distribution of inflammation to the prevertebral facia and surrounding tissues lead to laxity of ligaments and muscle contraction that it causes subluxation of atlanto-axial joint [3].

According to the previous research, the preexisting ligamentous laxity in children predisposes them to subluxation of first and second cervical vertebra. In addition, inflammatory process in prevertebral soft tissue surrounding first and second cervical vertebra can lead to subluxation of joint [4,2].

The diagnosis is based on plain X-ray, CT scan and MRI of neck. Increase in atlanto-axial distance may be suggestive for atlantoaxial subluxation. CT scan is most helpful method for establishing the diagnosis. MRI is useful for assessment of soft tissue inflammation [7].

The treatment includes immobilization of the neck, antibiotic therapy and muscle relaxants. The treatment should be started as soon as possible. Interruption in treatment may cause neck deformities that require surgical therapy and make irreversible neurological deficit [8].

Fielding’s classification assesses the degree of instability of first and second cervical vertebra subluxation. According to Fielding’s classification, type 1 and type 2 can be treated with conservative treatment while type III and type IV which are advanced stages need fixation and surgical intervention. In children extensive fusion should be avoided because of future skeleton growth [6].

Our case demonstrates that an inflammatory process in neck can lead to increased laxity of cervical ligaments leading to subluxation and Grisel’s syndrome.
Conflict of interest statement

We wish to confirm that there are no known conflicts of interest associated with this publication and there has been no significant financial support for this work that could have influenced its outcome.

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