Infectious process and torticollis, is there something to worry about?

Processos infecciosos e torcicolo, há algo com que se preocupar?

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RESUMO
A subluxação rotatória atlantoaxial em associação com processos infecciosos da cabeça e do pescoço (Síndrome de Grisel) é uma doença rara e pobremente relatada e não reportada. A patogênese da síndrome é incerta e diversas teorias foram postas para explicá-la. Este trabalho relata uma criança de 7 anos de idade com a Síndrome de Grisel e a literatura sobre o tema é brevemente revisada.

Palavras-chave: Infecção; Torcicolo; Síndrome de Grisel.

ABSTRACT
Atlantoaxial rotatory subluxation in association with head and neck infectious process (Grisel’s syndrome) is a rare disease and poorly reported. The syndrome’s pathogenesis is unclear and several theories have been proposed to explain it. This paper reports a 7 year old child with Grisel’s syndrome and the literature on this subject is briefly reviewed.

Keywords: Infection; Torticollis; Grisel’s syndrome.

INTRODUCTION
Atlantoaxial rotatory subluxation in association with head and neck infectious process (Grisel’s syndrome) is a rare disease. Most patients present with neck stiffness and torticollis. Patients usually are neurologically intact but neurological complications may occur in about 15% of the cases. The diagnosis may be underestimated or misdiagnosed due to the difficulties in making it. Early diagnosis is difficult and these patients without the correct management will be at risk of increasign atlantoaxial rotatory subluxation and neurological complications. The diagnosis is made by epidemiological correlation, clinical examination and radiological findings. Grisel’s syndrome is rare and poorly reported. We must keep in mind this diagnosis in patients with torticollis or cervical pain following an infectious process. This paper reports a 7 years-old child with Grisel’s syndrome and the literature on this subject is briefly reviewed.

CASE REPORT
A previously healthy 7 years-old boy was admitted to Neurological unit with a 50 days history of torticollis and cervical pain. The patient and his mother denied other symptoms or trauma. Two weeks before the symptoms began, he had a diagnosis of an upper respiratory tract infection with cervical lymphadenopathy. Clinical examination showed only a right torticollis and mild neck stiffness. Naso and oropharynx examination were normal and no other neurological impairment was found.

A cervical x-ray film showed a probable atlantoaxial subluxation (Figure 1). A cervical computed tomography (CT) revealed atlantoaxial rotatory subluxation with higher density of left cruciform ligament (Figure 1). A diagnosis of Grisel’s syndrome was made and the patient was immobilized with in a molded orthosis and received a 2-week course of analgesics with non steroidal anti-inflammatory...
Torticollis, a rotational deformity of the neck, is a common but nonspecific symptom in children. The differential diagnosis includes a large spectrum of diseases. Infections of the head and neck are a frequent cause, and the irritation with subsequent spasm of cervical or paraspinous muscles is generally benign and resolves with the control of infectious process. Atlantoaxial rotatory subluxation in association with head and neck infectious process is known as Grisel’s syndrome.\(^1\)

Atlantoaxial subluxation after head and neck infection has been rarely but consistently described in the literature since 1830, when Bell reported a patient with a syphilitic pharyngeal ulcer who died of acute spinal cord compression and an adolescent with a para-pharyngeal abscess who developed symptoms of spinal cord compression (postmortem examinations showed a C1–C2 dislocation). One hundred years later, Grisel, a French physician, reported on two cases of atlantoaxial subluxation occurring in children after pharyngitis and gave his name to the syndrome.\(^1,2,3,4,10\)

Grisel’s syndrome is predominant in children and is infrequent in adults. Cervical and occipital pain in addition to various neurological signs and symptoms are the usual clinical findings. Patients typically present with a characteristic head and neck posture termed a “cock-robin” deformity (reminiscent of a robin listening for worms) in which the chin is turned toward one side while the neck is laterally flexed to the opposite side.\(^7\) Most patients complain of neck stiffness and pain on attempted motion. There may be hypesthesia or hypalgia in the C2 dermatome, and sudden death has been reported.\(^3,9,10\) The main causes of Grisel’s syndrome are listed in Table 1.

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<thead>
<tr>
<th>Infection of head and neck</th>
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<td>Pharyngitis</td>
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<td>Adenoidectomy</td>
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<td>Adenotonsillitis</td>
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<td>Syphilitic pharyngeal ulcer</td>
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Table 1: Main infectious entities and otolaryngologic operations that have been associated with Grisel’s syndrome.

The rate of complications is directly proportional to the delay in the diagnosis and extent of complications depends on the degree of the subluxation and the severity of the cord compression. The duration of symptoms before syndrome identification varies from 2 days to 18 months with an average of 3 months.\(^2\)

The pathogenesis of the syndrome is unclear and several theories have been proposed to explain it. Most authors believed to...
be hyperemia following infection or surgical trauma that leads to decalcification of the anterior arch of the atlas and laxity of the anterior transverse ligament. There is a peri-odontoidal vascular plexus which drains the posterior superior pharyngeal region and with no lymph nodes, so septic exudates may be freely transverse from the pharynx to the atlantoaxial joint (this has been theorized as the anatomical rationale and cause for the syndrome). Knowledge of the biomechanics surrounding atlantoaxial rotatory subluxation is limited and it is possible that conditions affecting ligaments laxity (e.g., infections) or congenital atlantoaxial anatomic abnormalities increase the likelihood for atlantoaxial rotatory subluxation to occur.

Imaging exams confirm the diagnosis. CT scan, with and without three-dimensional (3D) reconstruction of the occipital bone and upper cervical spine, confirms Grisel’s syndrome. The diagnosis is confirmed radiographically when more than 2 mm of subluxation of the C1 lateral masses over C2. Cervical x-ray asymmetry of the lateral masses on anterior–posterior (AP) film is suggestive, but its interpretation may be difficult. MRI is helpful for soft tissue structures, effusion in the atlantoaxial articulation and can reveal occult infections in the upper respiratory tract.

The primary treatment is medical and the main goals of the therapy are bacteriologic cure, bony stability, and neurological protection. If early diagnosed, most cases require only non-operative treatment (eradication of infection, cervical traction until an adequate reduction and orthotic immobilization). When a proper diagnosis is delayed, intra and extra articular lesions may happen and hinder closed reduction of the dislocation. Surgical fusion is necessary for recurrent subluxation or for irreducible deformity.

Our patient was not treated with antibiotics because there were no signs of active infection in blood tests. Blood cultures were all negative and pharynx swab cultures showed normal flora. The use of symptomatics and orthosis were adequate for his recovery after 2 week with adequate reduction of atlantoaxial subluxation (Figure 2). The anterior transverse ligament was intact and an orthosis was sufficient. No neck pain, torticollis or recurrent subluxation were present on neurological examination 30, 60 and 90 days after discharge. He has full-range cervical mobility and he keeps on physical therapy.

References

Disclosure: The authors declare that they have no conflict of interest pertaining to this educational activity.

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