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CASE REPORT

Grisel's syndrome: A case report and review of the literature

Chad Galer [a,*], Eric Holbrook [b], John Treves [c], Donald Leopold [a]

[a] *Department of Otolaryngology, University of Nebraska Medical Center, 981225 Nebraska Medical Center, Omaha, NE 68198-1225, USA*
[b] *Massachusetts Eye and Ear Infirmary, Department of Otolaryngology, 981225 Nebraska Medical Center, Omaha, NE 68198-1225, USA*
[c] *Section of Neurosurgery, University of Nebraska Medical Center, 981225 Nebraska Medical Center, Omaha, NE 68198-1225, USA*

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Summary Grisel's syndrome is non-traumatic atlantoaxial subluxation (AAS) secondary to an inflammatory process in the upper neck. It is a rare condition that occurs almost exclusively in children and has been associated with upper cervical infections and otolaryngologic procedures. A case of AAS secondary to an upper cervical infection is presented. Potential sequelae can be severe; early diagnosis and treatment of Grisel's syndrome can prevent tragic outcome.

1. Introduction

Grisel's syndrome is non-traumatic atlantoaxial subluxation (AAS) secondary to an inflammatory process in the upper neck. It was first reported by Bell in 1830, but is named after Grisel who published on two cases in 1930 [1]. Grisel's is a rare condition that occurs almost exclusively in children and has been associated with upper cervical infections and otolaryngologic procedures. Few cases of this rare syndrome have been reported in the last 35 years. We present the case of a 3-year old girl with Grisel's syndrome caused by cervical infection.

2. Case report

A 3-year old girl was referred for evaluation of sore throat, neck pain, and fever. She had been seen in the Emergency Department 3 days previously, was diagnosed with tonsillitis and started on a course of oral antibiotics. In the interim, she continued to have fevers and sore throat. Her past medical history was non-contributory. At physical examination, the patient was noted to hold her head slightly rotated and bent to the left. Computed tomography of the neck with contrast was obtained which revealed a 2.1 cm × 1.1 cm × 3 cm low density fluid collection in the prevertebral space superior and posterior to the left palatine tonsil (Fig. 1). Due to the location of the lesion and the patients good overall status, we elected to treat conservatively

* Corresponding author. Tel.: +1 402 559 8007; fax: +1 402 559 8940.
E-mail address: cgaler@unmc.edu (C. Galer).

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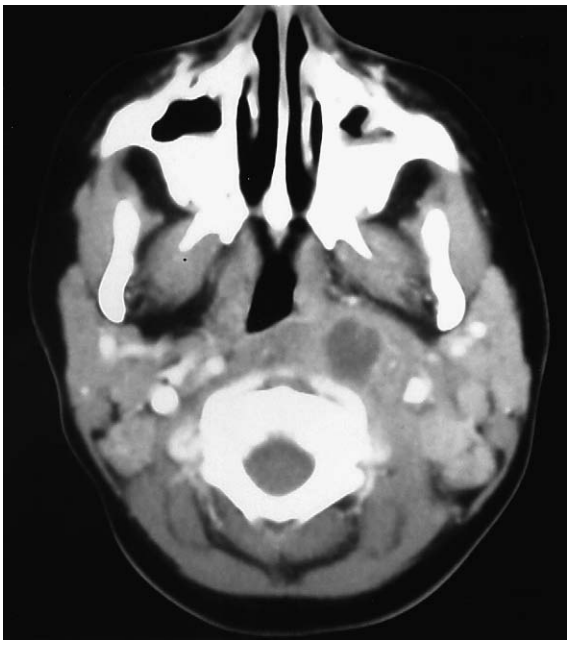

Fig. 1 CT scan showing a 2.1 cm × 1.1 cm × 3 cm low density fluid collection in the prevertebral space. No subluxation is evident.

with intravenous antibiotics. A repeat CT scan of the neck was obtained on hospital day 3 and showed persistence of the fluid collection as well as a C1-C2 atlantoaxial subluxation (Fig. 2). At this point, the neurosurgical service was consulted. The patient was taken to the operating room for peroral needle drainage of the fluid collection and reduction of the subluxation by cervical traction and placement of a hard cervical collar. Attempts to aspirate the collection were unsuccessful. The subluxation was well reduced and the child was sent home on the subsequent day with cervical collar and 4 weeks of intravenous antibiotics. Repeat CT scan one month later revealed normal alignment of the cervical vertebrae as well as resolution of the fluid collection (Fig. 3).

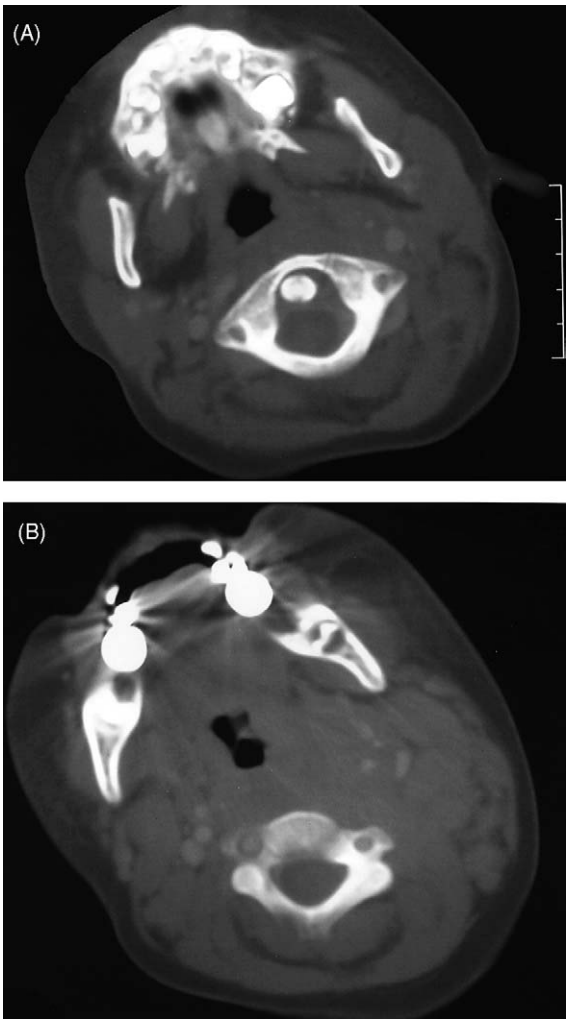

Fig. 2 CT scan obtained 3 days later shows persistence of the fluid collection. Rotary subluxation is demonstrated by the orientation of the bodies of C1 (A) and C2 (B) and the eccentric position of the odontoid process in relation to the C1 arch (A).

3. Discussion

The exact etiology of Grisel's syndrome is controversial. It is generally accepted that the process is initiated by an infection in the upper cervical region which creates an inflammatory milieu. Grisel postulated that AAS was caused by muscle spasm; now muscle spasm is thought to be a reaction to adjacent inflammation and the role of this muscle spasm is limited to maintenance of the subluxed position but is not causative [1,2]. Hyperemia of the blood supply of bone has been shown to cause resorption. Watson-Jones suggested that resorption of the attachment of the transverse ligament to the atlas mediated by inflammation could lead to AAS. In a case report by Watson-Jones and Roberts, a patient with Grisel's syndrome was found to have a decalcified anterior arch of the atlas, which reappeared after reduction and treatment of the inciting infection [3]. However, this was not substantiated in a meta-analysis of post-mortem specimens and roentgenograms of patients with Grisel's syndrome by Pandya [4]. Wittek suggested that the etiology may be due to distension of the ligaments with maintenance of the bony-ligamentous attachment [5]. This is supported by experiments in rabbit knee joints that have shown that a sterile inflammatory environment causes marked weakness and laxity of the surrounding ligaments [6]. More recently, Welinder et al. [7] provided radiologic evidence for this