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Clinical Record

Grisel syndrome: a delayed presentation in an asymptomatic patient

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Abstract

Grisel syndrome is a rare condition characterised by atlanto-axial subluxation following an inflammatory process in the head and neck region. It occurs more commonly in children and usually presents with cervical pain and torticollis, in addition to symptoms of the primary infection. We present the case of an asymptomatic 78-year-old man who was incidentally found to have atlanto-axial subluxation on a routine follow-up computed tomography scan, three months following successful treatment of a skull base infection. This case emphasises the importance of appropriate follow-up imaging for patients with skull base infections, even if they respond clinically to medical treatment.

Key words: Atlanto-Axial Joint; Skull Base; Infection

Introduction

Grisel syndrome is a rare condition characterised by atlanto-axial subluxation following an inflammatory process in the head and neck region. It has been described following otolaryngological procedures such as tonsillectomy, mastoid surgery and repair of choanal atresia, and also after infections of the head and neck.[1,2] It predominately affects children, although cases in adults have been described.[3] It usually presents with cervical pain and torticollis, in addition to symptoms of the primary infection.

We present a case report of an asymptomatic adult who was incidentally diagnosed with atlanto-axial subluxation on follow-up imaging, three months after successful treatment of a skull base infection.

Case report

A 78-year-old man presented to the out-patient clinic with a three-week history of hoarse voice, dysphagia, weight loss, a discharging left ear, otalgia and neck pain. There were no symptoms of tinnitus or hearing loss. Past medical history included tablet-treated diabetes mellitus and hypertension, which were well controlled. He had no alcohol or smoking history.

On examination, the left external auditory canal was moist, consistent with active otitis externa. The tympanic membrane was slightly injected, with no evidence of attic or external auditory canal polyps or granulations. Positive findings on cranial nerve examination included wasting of the left side of the tongue and a left facial nerve palsy (House–Brackmann grade two) which, according to the patient, was long-standing. Flexible nasendoscopy revealed a normal post-nasal space and a left vocal fold palsy, with a maximum phonation time of five seconds. The cervical spine was tender on palpation and torticollis to the left was present, with no evidence of meningism or peripheral neurology. Blood tests revealed a normal white cell count and a raised C-reactive protein at 118 mg/l.

The patient was commenced on topical aural Sofradex® and intravenous Augmentin®, in addition to analgesia, for a presumed malignant otitis externa with cranial nerve involvement. However, a computed tomography (CT) scan revealed a 3.3 × 3 cm, soft tissue mass below the skull base, extending onto the lateral pterygoid, with erosion of the left occipital condyle, jugular foramen and hypoglossal canal (Figure 1), consistent with non-otogenic skull base osteomyelitis. An ear swab was negative for pseudomonas species. After discussion with the microbiologist, the patient was continued on intravenous Augmentin. A white cell scan showed uptake at the left base of skull region, further suggesting an inflammatory process rather than a malignancy. Biopsies from the post-nasal space showed no evidence of malignancy.

Long-term intravenous antibiotics were continued via a Hickman line, and the patient improved clinically, with a corresponding decrease in inflammatory marker levels. He left hospital, after six weeks of intravenous antibiotics, with normal inflammatory blood markers on discharge.

A repeat CT scan at three months following discharge was performed to reassess the inflammatory mass seen on the original scan. This scan revealed a marked reduction in the size of the left skull base inflammatory mass, but also noted was an atlanto-axial subluxation of 1.2 cm, with superior displacement of the odontoid peg (Figure 2). This had not been present on the original CT scan.

The patient was referred to the neurosurgeons, who successfully performed an open reduction and internal fixation of the subluxated atlanto-axial joint (Figure 3). Follow-up assessment, one month following the patient's spinal surgery, was unremarkable.

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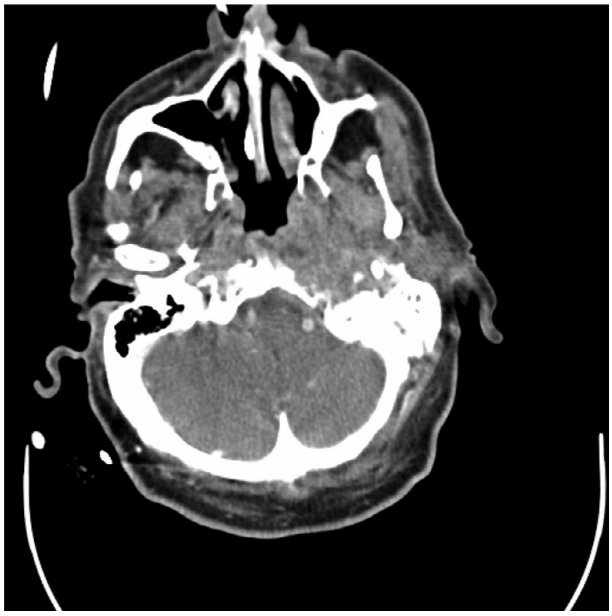

FIG. 1
Axial computed tomography scan showing left skull base inflammatory mass.

Discussion

The exact cause of the atlanto-axial subluxation found in Grisel syndrome is not fully understood, but it involves disruption of the ligamentous support of the atlanto-axial joint. Several theories have been proposed.[1] Oedema of the atlanto-axial ligaments and a consequent increase in laxity may spread infection from pharyngovertebral veins. These veins drain from the posterior pharyngeal region and are in close proximity to the atlanto-axial joint. According to this theory, the torticollis which accompanies Grisel syndrome is a protective mechanism from overlying muscles trying to stabilise the lax atlanto-axial joint.

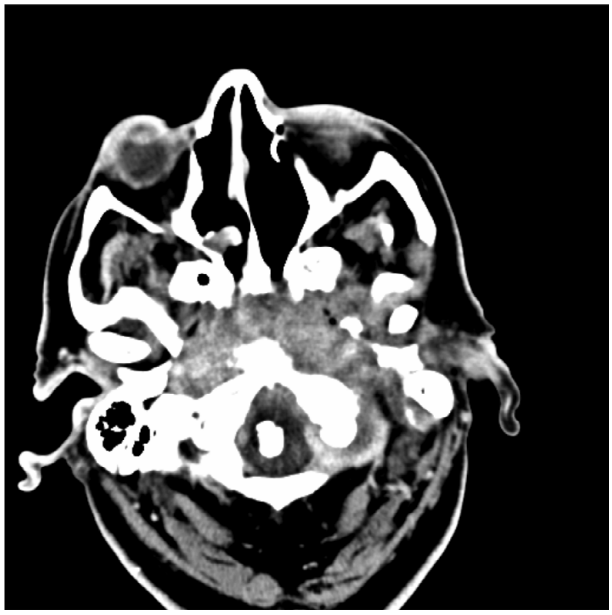

FIG. 2
Axial computed tomography scan showing atlanto-axial subluxation.

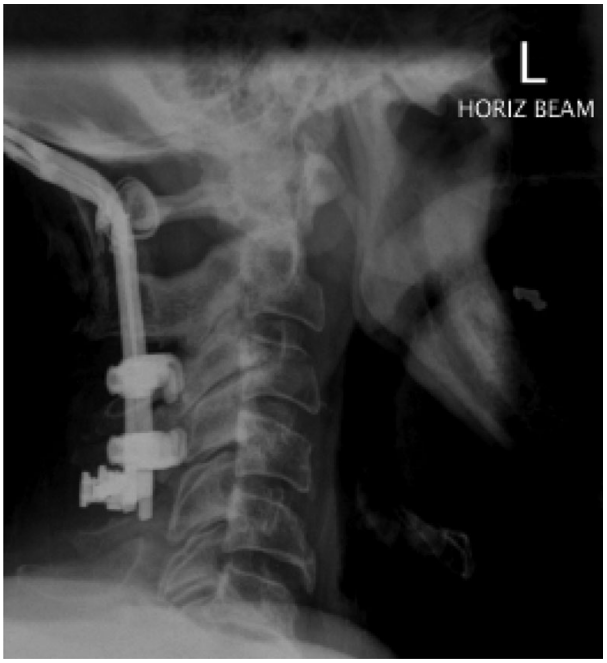

FIG. 3
Plain radiograph showing surgical stabilisation of atlanto-axial subluxation. L = left; Horiz = Horizontal

An alternative theory is that the torticollis causes excessive traction on the atlanto-axial ligaments and is the primary cause of the subluxation, rather than being a protective mechanism.

The clinical findings in cases of atlanto-axial subluxation are variable. The neck is usually tender and the torticollis is opposite to the side of subluxation. The spinous process of the axis opposite to the side of the subluxation may be palpable (Sudek's sign). The presence of neurological sequelae depends upon the severity of the subluxation, which can be graded using the Fielding classification (Table I).[4]

The investigation of choice is either a high resolution CT scan to demonstrate bony subluxation or a magnetic resonance imaging scan to demonstrate inflammation of the atlanto-axial ligaments.[2] Plain radiographs may demonstrate asymmetry between the facet joints and some decalcification; however, appearances can be non-diagnostic. Lateral cervical spine radiographs (flexion and extension) may show atlanto-axial subluxation.

Management of Grisel syndrome includes treatment of the underlying infection and an orthopaedic or neurosurgical opinion of the atlanto-axial subluxation. Depending on the severity of the subluxation, treatment can include use of a soft collar, cervical traction or surgical stabilisation.

TABLE I
FIELDING CLASSIFICATION OF ATLANTO-AXIAL SUBLUXATION

Type	Atlanto-axial joint displacement
I	Rotary fixation without displacement
II	Rotary fixation with anterior displacement of atlas (3–5 mm)
III	Rotary fixation with anterior displacement of atlas (>5 mm)
IV	Rotary fixation with posterior displacement of atlas

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In our case, the initial diagnosis of malignant otitis externa was reasonable in view of the active ear infection and cranial nerve palsies. Sofradex and Augmentin were commenced, as our department does not normally begin anti-pseudomonal treatment until a positive microbiology culture and sensitivity report have been obtained. However, due to the absence of a pseudomonas species growth from the ear, the absence of aural granulations or polyps, and the CT scan result, the diagnosis was changed to non-otogenic skull base infection. The CT scan performed three months after the clinical resolution of the infection showed an incidental, grade three atlanto-axial subluxation.

- **Grisel syndrome is a rare condition characterised by atlanto-axial subluxation following an inflammatory process in the head and neck region**
- **It usually presents with symptoms or signs of atlanto-axial subluxation, in association with the primary inflammatory process**
- **It can have devastating consequences if not detected and treated early**
- **Grisel syndrome can present after a delayed interval, several months after successful treatment of the inflammatory process**
- **Follow-up imaging is important to detect atlanto-axial subluxation, which can occur in an asymptomatic patient**

This case demonstrates that this rare condition can occur months following the initial infection, despite successful treatment, and can present in an asymptomatic individual. This highlights the importance of follow-up imaging in patients who initially present with signs and symptoms suggestive of atlanto-axial involvement (i.e. neck pain, torticollis and neck tenderness), even if they become asymptomatic following medical treatment of their initial infection or inflammation.

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