

Grisel's Syndrome: Non-Traumatic Subluxation of the Atlantoaxial Joint

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A non-traumatic subluxation of the atlantoaxial joint in association with inflammation in the head and neck region is known in neurosurgery as Grisel's syndrome¹. The inflammation usually results from an infection, either systemic via hematogenous spread or regional, by contiguity, as a complication of head and neck surgery. It leads to laxity of the ligaments supporting the atlantoaxial joint, and subluxation. This uncommon entity is a pediatric disease but rare adult cases do occur. Patients present with very painful torticollis and fever. If left untreated, neurological complications in the form of radiculopathy, quadriplegia, and even death have been estimated to occur in 15% of cases². Early diagnosis is therefore of great importance and fortunately modern imaging has made it possible to confirm the diagnosis much earlier and prevent late consequences of Grisel's syndrome³.

A previously healthy 51-year-old man presented with a 2-day history of excruciating neck pain, torticollis, and fever. Two days earlier, he had suffered a severe ankle sprain while

playing squash. On the following day, neck pain developed and despite the use of analgesics, the pain became increasingly more intense over the ensuing days. There was no other history of trauma. Physical examination revealed a temperature of 38.0°C, severe pain with any neck movement, and left ankle synovitis. There was no neurological impairment. The white blood cell count was 8.0 with a normal differential, and the C-reactive protein level was 296 mg/l (normal < 5 mg/l). Blood, urine, and left ankle synovial fluid cultures all grew *Staphylococcus aureus*. Magnetic resonance imaging (MRI) scan of the C-spine, performed at presentation, showed an inflammatory/infectious process at the atlantodental space which extended to involve the pre-vertebral soft tissues (Figure 1). Early diagnosis resulted in prompt initiation of antibiotics. Recovery was clinically complete and without sequelae on repeat MRI.

This patient's condition would now be more accurately referred to as "pre-Grisel's syndrome," an inflammation around the atlantoaxial joint associated with torticollis, prior to the development of subluxation and consequent neurological complications.

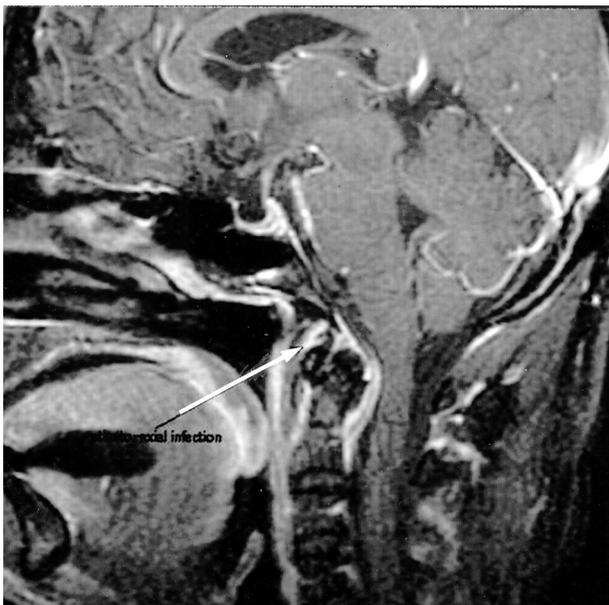


Figure 1. Sagittal T1-weighted magnetic resonance image with fat saturation and gadolinium enhancement showing robust soft tissue enhancement around the atlantoaxial joint and perivertebral tissues (arrow).

REFERENCES

1. Gourin CG, Kaper B, Abdu WA, Donegan JO. Nontraumatic atlanto-axial subluxation after retropharyngeal cellulitis: Grisel's syndrome. *Am J Otolaryngol* 2002;23:60-5.
2. Yu K, White DR, Weissler MC, Pillsbury HC. Nontraumatic atlantoaxial subluxation (Grisel syndrome): a rare complication of otolaryngological procedures. *Laryngoscope* 2003;113:1047-9.
3. Ugur HC, Çağlar S, Unlu A, Erdem A, Kanpolat Y. Infection-related atlantoaxial subluxation in two adults: Grisel syndrome or not? *Acta Neurochir* 2003;145:69-72.