Turk J Phys Med Rehab 2021;67(2):259-263
DOI: 10.5606/tftrd.2021.4967
Available online at www.turkishjournalpmr.com

Case Report

# Unusual presentation of an unusual disease: A very delayed diagnosis of Grisel's syndrome

Cevriye Mülkoğlu, Hakan Genç, Seçil Vural, Başak Mansız-Kaplan

Department of Physical Medicine and Rehabilitation, Ankara Training and Research Hospital, Ankara, Turkey

**Received:** June 25, 2019 **Accepted:** December 26, 2019 **Published online:** May 25, 2021

***ABSTRACT***

Grisel's syndrome (GS) is a rare syndrome which refers only to non-traumatic atlantoaxial subluxation. This syndrome predominantly occurs in young children following an upper respiratory infection or otolaryngologic procedures. An eight-year-old girl with a delayed diagnosis of GS was admitted to our outpatient clinic with complaints of painful torticollis and neck stiffness. Three-dimensional computed tomography revealed rotatory atlantoaxial subluxation. After consulting with the neurosurgery department, the patient underwent surgery. The significance of this patient was that she was unable to be diagnosed early and atlantoaxial subluxation remained hidden for five years without any complications. In conclusion, this rare case highlights the importance of delayed diagnosis of GS and clinicians should be aware of this syndrome.

***Keywords:*** Grisel's syndrome, neck stiffness, non-traumatic atlantoaxial subluxation, torticollis.

Grisel's syndrome (GS) is characterized by atlantoaxial joint subluxation without trauma or bone disease.[1] This uncommon syndrome usually appears in young children and, rarely, in adults.[2] It occurs secondary to head and neck infections, nasopharyngeal inflammation, or recent ear, nose and throat surgery.[1,3] The pathogenesis of this syndrome still remains unclear. Although several theories have been discussed so far, the most accepted is the increased ligament flexibility during inflammation of head and neck, which is the primary reason for atlantoaxial subluxation.[4,5] Patients usually present with painful torticollis and restricted cervical movements. The diagnosis of GS is based on clinical examination, medical history, and strong suspicion of this syndrome. Early diagnosis and treatment are critical factors for a good prognosis.[2,3,6]

In this report, we present a rare case of delayed diagnosis of GS admitted to our clinic with complaints of neck pain and torticollis.

## CASE REPORT

An eight-year-old girl was referred to our outpatient clinic with complaints of neck asymmetry, restricted cervical movements, and pain on the right side of her neck. Her parents noticed an asymmetry on her neck at three years of age. She had no history of head and neck trauma. She had no history of upper cervical or head infection, or otolaryngologic procedure, either. She had painful torticollis which was misdiagnosed during multiple visits to the pediatric, neurosurgical, and orthopedic departments. Initially, it was thought that this condition was a muscular torticollis and stretching and strengthening exercises were recommended by previous physicians from different specialties. On her physical examination, the throat was normal. She had no fever. Her head was in the cock-robin position (Figure 1); i.e., tilted toward the affected side and flexed anteriorly, and the chin was turned to the opposite side.[4] Torticollis and shortness of the right sternocleidomastoid muscle (SCM) were

**Corresponding author:** Cevriye Mülkoğlu, MD. Ankara Eğitim ve Araştırma Hastanesi Fizik Tedavi ve Rehabilitasyon Kliniği, 06230 Altındağ, Ankara, Türkiye.
e-mail: drckaraca@hotmail.com

*Cite this article as:*
Mülkoğlu C, Genç H, Vural S, Mansız-Kaplan B. Unusual presentation of an unusual disease: A very delayed diagnosis of Grisel's syndrome. Turk J Phys Med Rehab 2021;67(2):259-263.

**Figure 1.** There was a cock-robin posture in inspection, with a tilted neck toward to the affected side and turned chin to the opposite side.

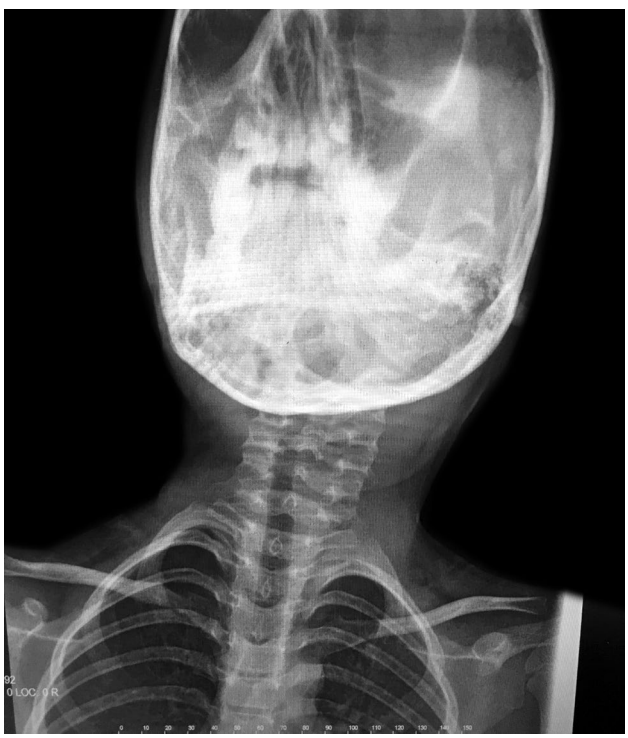

**Figure 2.** Cock-robin position on anterior-posterior direct cervicography.

observed on inspection. Cervical lateral flexion and cervical rotation to the left and cervical extension were restricted by approximately 20°. Her neurological examination findings were normal. The results of all biochemical blood tests were within normal ranges. The cock-robin position was visualized on the anteroposterior cervical plain graph (Figure 2). The three-dimensional (3D) computed tomography (CT) revealed atlantoaxial rotatory subluxation at an angle of 10° on the transverse plane (Figure 3). Additionally, cervical magnetic resonance imaging (MRI) confirmed the atlantoaxial rotatory subluxation without any soft tissue pathology. Based on these clinical and radiological findings, the diagnosis of GS was made. The patient was consulted to neurosurgery department and resection myotomy on the right SCM was decided. Atlantoaxial joint surgery was not considered due to the fixed rotatory subluxation persisting without any neurological complication. A written informed consent was obtained from each parent. A resection myotomy was successfully performed and the patient was discharged with a cervical orthosis on the fifth day of the surgery.

## DISCUSSION

Grisel's syndrome is characterized by non-traumatic subluxation of the joint between the atlas and axis.[7,8] In 1951, Grisel,[9] who gave his name to this syndrome, reported two cases in which atlantoaxial subluxation developed after pharyngitis. Although GS typically appears in young children,

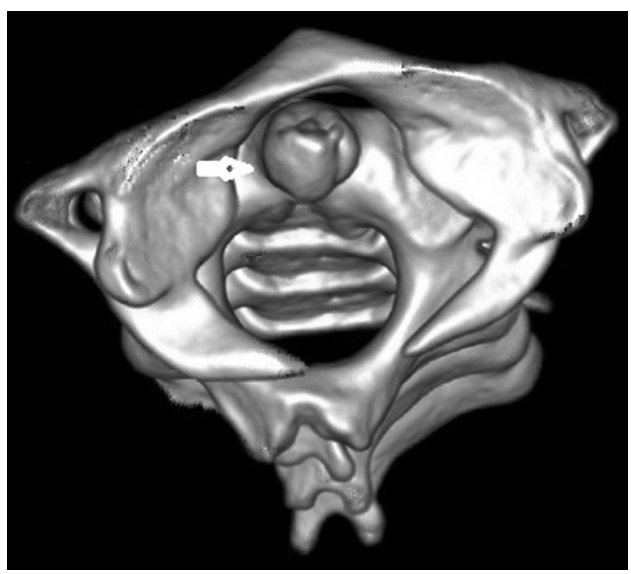

**Figure 3.** Three-dimensional computed tomography revealed a rotatory atlantoaxial subluxation.

particularly between ages of 5 to 12 years, adult cases can be also seen.[1,2]

Several entities have been discussed in the literature to be associated with GS including pharyngitis, nasopharyngitis, adenotonsillitis, tonsillar and cervical abscesses, parotitis, and otitis media and otolaryngologic procedures such as tonsillectomy, adenoidectomy, mastoidectomy, choanal atresia repair, and cleft palate repair.[2,4,9] Karkos et al.[10] reported 96 cases with non-traumatic atlantoaxial rotary subluxation: 48% due to infections and 40% after ear, nose and throat surgery (adenotonsillectomy in 78%). The infectious etiology included upper respiratory tract viral infections (83%), retropharyngeal abscesses (11%), otitis media (4%), and mumps (2%). However, none of these associated conditions was present in our patient. The most common reason for GS is infections; however, the infection may be insignificant or distant from the head and neck region and, therefore, it can be unrecognized or overlooked by the parents. In this patient, the diagnosis was delayed for five years, as the parents did not describe a definite infection or operation about the upper respiratory tract.

Grisel's syndrome may frequently affect patients with congenital ligamentous laxity, such as Down syndrome and Marfan syndrome. Torticollis may occur due to many different reasons. Congenital muscular torticollis, hypertrophy or absence of cervical muscles, spina bifida, posterior fossa and spinal cord tumors, cerebral palsy, Arnold-Chiari malformation, Klippel-Feil syndrome, syringomyelia, malignancy, fractures or congenital anomalies of cervical vertebrae should be considered among the differential diagnosis of torticollis.[5,11] Our patient had no congenital or rheumatologic disease.

Although several theories have been discussed to explain the pathogenesis of GS, the widely accepted theory is hematogenous spreading of infections from the posterior pharynx to the upper cervical joints with hyperemia and abnormal relaxation of the transverse ligament.[12] Anatomical data support this theory. Since the periodontoidal venous plexus drains the posterior superior pharyngeal region and has no lymph nodes, septic inflammation may easily spread from the pharynx to the upper cervical joints.[1,2,12] The main stabilizer of the atlantoaxial joint is the transverse ligament, while the secondary stabilizers are alar ligaments.[2] Nasopharyngeal infection and inflammation cause hyperemia that may weaken the ligaments between the atlas and axis, resulting in subluxation.[9,12]

The diagnosis of GS is based on clinical examination and imaging studies. Neck pain and painful torticollis are significant symptoms. Lhermitte's sign may be also present in some cases.[4] The main three indications of GS are Sudeck's sign, consisting of the palpable deviation of the spinous process of the axis in the same direction of the head rotation, the cock-robin sign, which refers to torticollis with the chin turned away from the subluxated side and the head tilted toward the subluxed side and flexed anteriorly, causing the patient to have difficulty in opening his/her mouth, and spasm of ipsilateral SCM, in which restricted cervical movements may lead to dysphagia, trismus, hearing loss, and craniofacial asymmetry. Radiological imaging is a valuable tool in the diagnosis and follow-up of patients with GS. The anteroposterior plain graph shows asymmetry between the facet joints. Lateral graphs can show an increased atlantodental interval. The normal atlantodental distance is ≤3 mm in adults and ≤5 mm in children. However, normal evaluation findings on plain radiograph does not necessarily rule out atlantoaxial subluxation.[1,2] The 3D CT is the gold standard for the assessment of the atlantoaxial joint.[5] In addition, MRI reveals retropharyngeal soft tissue swelling, as well as spinal cord compression.[3,5] Neurological complications occur in nearly 15% of cases and lead to radiculopathy, myelopathy, quadriplegia, and death from cord compression.[1,2,5,7] Serious sequelae can be prevented, if the diagnosis and treatment of GS is undertaken earlier.[2,12]

There are four types of subluxation according to Fielding and Hawkins classification.[13] Type I shows a fixed rotation of atlas and axis without subluxation, anterior dislocation of the atlas less than 3 mm; the transverse ligament is intact. In Type II, ventral dislocation of the atlas on unilateral articular process with 3 to 5-mm displacement. Type III consists of rotation of the atlas with ventral displacement in both joints more than 5 mm. In Type IV, there was a rotatory fixation and posterior subluxation of the atlas. Combination with fractured dens axis or congenital dens-aplasia is possible. Our patient was compatible with Type II dislocation.

The first-line treatment of GS should be conservative including bed rest, antibiotic therapy, muscle relaxants, anti-inflammatory therapy, and neck immobilization with soft or hard collars, cervical traction, and external fixation. A treatment regimen for atlantoaxial subluxation has been proposed according to the Fielding and Hawkins

classification.[13-16] Conservative management provides spontaneous reduction in majority of Type I, II, and III cases.[10] In the treatment of Type I, bed rest, muscle relaxants, non-steroid anti-inflammatory agents, and immobilization with a soft cervical collar are usually sufficient. Type II subluxation requires also conservative treatment, reduction, and cervical traction with a stiff cervical collar (Philadelphia™ collar [BSN Medical Inc., NC, USA] or sterno-occipital-mandibular immobilization device [SOMI] brace). Type III requires closed reposition and halo-extension. Following six to eight weeks of external immobilization, cervical stability is assessed by the flexion-extension radiographs. Holcomb et al.[17] recommended that once the reduction was radiologically confirmed, a halo vest should be applied for 6 to 12 weeks. If residual instability is indicated, C1-C2 arthrodesis is proposed.[14] Type III and IV subluxations usually need bed rest in cervical traction, followed by a period of neck immobilization in a cervical collar to prevent recurrent subluxation. In Type IV, C1-C2 fusion is recommended for treatment of subluxation.

In most of the cases, head halter traction is appropriately used, while Gardner-Wells tongs may be required in the long-standing and severe deformities.[15] External immobilization is usually used for six to eight weeks, following which flexion-extension radiographs are needed in deciding any residual subluxation. If there is an evidence of residual subluxation, arthrodesis is indicated.[14,17] Surgery is considered for only patients with failed conservative treatment having recurrent or irreducible subluxation.[15] Surgical interventions include open reduction of the deformity and fusion of the C1-2 vertebrae with a bone graft.[3,15]

Fielding and Hawkins[13] recommended surgical fusion of C1-C2 in cases with a delayed diagnosis (of more than three months) to avoid recurrent subluxation. Of note, GS may occur even months after local infections. If the diagnosis is established within four weeks, manual reposition seems to be adequate. In patients with a delayed diagnosis, many authors recommend immobilization with halo-fixation for 12 weeks.[13,16]

In a case report, Pilge et al.[16] reported a pediatric case with torticollis after insertion of a cochlea implant treated with manual repositioning under general anesthesia and temporary immobilization with a cervical collar for two weeks. The authors achieved good repositioning within a short period of time. Similarly, Akbay et al.[18] described a closed manual reduction maneuver to reduce Type I and II atlantoaxial rotatory subluxation in children. The patients used a SOMI brace after discharge for four weeks. In 11 of 12 pediatric patients, dislocation was successfully reducted with closed manual reduction.[18] In the series (n=4) of Fernández Cornejo,[19] none of the patients needed surgery; however, in one patient, persistent restriction of the neck developed. In another study of Deichmueller et al.,[20] eight of 12 patients had full recovery following intravenous antibiotics and oral anti-inflammatory therapies alone. Four patients required reduction of the atlantoaxial joint under general anesthesia and external immobilization by halo-fixation for six weeks. Our patient was referred to our clinic at the chronic phase. The inflammation in the region of the atlantoaxial joint healed spontaneously and atlantoaxial rotatory subluxation became fixed with the fibrous tissue. Traction, manual reduction, external immobilization or atlantoaxial surgery were not considered for the present case, due to the long-lasting fixed rotatory subluxation without any neurological complications.

In conclusion, conservative treatment for GS usually improves the symptoms. Clinicians should be aware of GS in patients who complain about neck pain and persistent torticollis, particularly after upper respiratory tract infections.

**Declaration of conflicting interests**

The authors declared no conflicts of interest with respect to the authorship and/or publication of this article.

**Funding**

The authors received no financial support for the research and/or authorship of this article.

## REFERENCES

1. Bucak A, Ulu S, Aycicek A, Kacar E, Miman MC. Grisel's Syndrome: A Rare Complication following Adenotonsillectomy. Case Rep Otolaryngol 2014;2014:703021.
2. Ugur HC, Cağlar S, Unlu A, Erdem A, Kanpolat Y. Infection-related atlantoaxial subluxation in two adults: Grisel syndrome or not? Acta Neurochir (Wien) 2003;145:69-72.
3. Spennato P, Nicosia G, Rapanà A, Cicala D, Donnianni T, Scala S, et al. Grisel Syndrome Following Adenoidectomy: Surgical Management in a Case with Delayed Diagnosis. World Neurosurg 2015;84:1494.e7-12.
4. Youssef K, Daniel S. Grisel syndrome in adult patients. Report of two cases and review of the literature. Can J Neurol Sci 2009;36:109-13.
5. Cellucci T, Tse SM, Shroff M. Case 2: A pain in the neck. Paediatr Child Health 2012;17:247-50.

6. Aladag Ciftdemir N, Eren T, Ciftdemir M. A Rare Cause of Torticollis: Grisel Syndrome. J Trop Pediatr 2018;64:245-8.
7. Nozaki F, Kusunoki T, Tomoda Y, Hiejima I, Hayashi A, Kumada T, et al. Grisel syndrome as a complication of Kawasaki disease: a case report and review of the literature. Eur J Pediatr 2013;172:119-21.
8. Fath L, Cebula H, Santin MN, Coca A, Debry C, Proust F. The Grisel's syndrome: A non-traumatic subluxation of the atlantoaxial joint. Neurochirurgie 2018;64:327-30.
9. Gupta A, Prakash J, Kumar P, Singh NN. Anesthetic issues and Difficult Airway Management in a Case of Grisel's Syndrome. Anesth Essays Res 2017;11:1094-6.
10. Karkos PD, Benton J, Leong SC, Mushi E, Sivaji N, Assimakopoulos DA. Grisel's syndrome in otolaryngology: a systematic review. Int J Pediatr Otorhinolaryngol 2007;71:1823-7.
11. Gray GM, Tasso KH. Differential diagnosis of torticollis: a case report. Pediatr Phys Ther 2009;21:369-74.
12. Ahn AR, Park YH, Park EJ, Yim SY. A Case of Grisel Syndrome Showing No Underlying Laxity of the Atlanto-axial Joint. Ann Rehabil Med 2017;41:511-5.
13. Fielding JW, Hawkins RJ. Atlanto-axial rotatory fixation. (Fixed rotatory subluxation of the atlanto-axial joint). J Bone Joint Surg [Am] 1977;59:37-44.
14. Wetzel FT, La Rocca H. Grisel's syndrome. Clin Orthop Relat Res 1989;(240):141-52.
15. Rinaldo A, Mondin V, Suárez C, Genden EM, Ferlito A. Grisel's syndrome in head and neck practice. Oral Oncol 2005;41:966-70.
16. Pilge H, Prodinger PM, Bürklein D, Holzapfel BM, Lauen J. Nontraumatic subluxation of the atlanto-axial joint as rare form of acquired torticollis: diagnosis and clinical features of the Grisel's syndrome. Spine (Phila Pa 1976) 2011;36:E747-51.
17. Holcomb JD, Jaffe DM, Greinwald JH Jr, Bauman NM, Smith RJ. Nontraumatic atlantoaxial rotary subluxation in the pediatric otolaryngology patient. A report of four cases. Ann Otol Rhinol Laryngol 2001;110:1137-40.
18. Akbay A, Bilginer B, Akalan N. Closed manual reduction maneuver of atlantoaxial rotatory dislocation in pediatric age. Childs Nerv Syst 2014;30:1083-9.
19. Fernández Cornejo VJ, Martínez-Lage JF, Piqueras C, Gelabert A, Poza M. Inflammatory atlanto-axial subluxation (Grisel's syndrome) in children: clinical diagnosis and management. Childs Nerv Syst 2003;19:342-7.
20. Deichmueller CM, Welkoborsky HJ. Grisel's syndrome--a rare complication following "small" operations and infections in the ENT region. Eur Arch Otorhinolaryngol 2010;267:1467-73.