



Early diagnosis of Grisel's syndrome in children with favorable outcome

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Abstract

Purpose The painful torticollis only itself may be a major sign for the diagnosis of Grisel's syndrome (GS). It is known as an inflammatory atlantoaxial rotatory subluxation following upper respiratory tract infections (URTI) and surgical otolaryngological procedures.

Patients and methods The analysis of 16 pediatric GS patients were reviewed retrospectively by considering the diagnosis, the treatment modality, and the prognosis at the Department of Neurosurgery and Otorhinolaryngology in Mersin University, Faculty of Medicine between 2008 and 2018. In addition to the clinical cases, five cadavers were used to demonstrate atlantoaxial region, particularly the ligamentous complex and articulation of the atlas-axis, for the mechanism of these rare entities.

Results The most common etiological factor of GS was URTI with 81.25% among 16 patients. Painful torticollis was the primary symptom of pediatric patients at admission. The X-Ray, computerized tomography (CT), and magnetic resonance imaging (MRI) investigations were used for the definitive diagnosis in the first week of admission, except one. No morbidity, mortality, and deformity were reported in this series.

Conclusions Early diagnosis is the principle of GS for avoiding of permanent neck deformity and complex surgical procedures. If GS can be diagnosed without any doubt by only considering patient's history and clinical examination, CT scan is not recommended due to harmful effects of radiation. The treatment was achieved by reduction, external fixation under analgesia, or sedoanalgesia accompanying with antibiotic and anti-phylogistic treatment.

Keywords Atlantoaxial rotatory subluxation · Grisel's syndrome · Painful torticollis · Upper respiratory tract infection · Pediatric patient

Introduction

Grisel's syndrome (GS) is one of the remarkable entities that should be considered for painful torticollis in especially pediatric cases. It is usually known as non-traumatic inflammatory atlantoaxial rotatory subluxation after upper respiratory tract infections (URTI) such as pharyngitis,

adenotonsillitis, tonsillar abscesses, cervical abscesses, otitis media, and various otolaryngological surgical interventions including tonsillectomy, adenoidectomy, and mastoidectomy [1–7]. The symptomology of these patients can be listed as cervical pain, head tilt, and limited neck movement resulting with torticollis.

GS was first described by Bell in one of his autopsy cases that the transverse ligament had been eroded due to syphilitic ulceration of pharynx in 1830 [8]. Furthermore, the name of Grisel is originated from the French physician, otolaryngologist Pierre Grisel whom described this syndrome in two of the nasopharyngitis patients in 1930 [9].

The retropharyngeal space provides an effective barrier against vascular connections between prevertebral fascia and peripharyngeal tissue. However, the pharyngovertebral veins go through the periodontoid venous plexus and finally drain into the upper cervical epidural sinuses. Meanwhile, the odontoid process is surrounded by terminal branches of ascending pharyngeal artery and vascular arcades that were the

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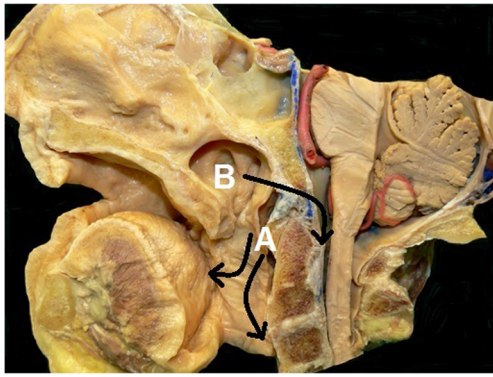


Fig. 1 Periodontoidal and suboccipital venous plexus relationship between pharyngovertebral area has been demonstrated which serves a septic hemotogenous route for the upper cervical spine region for the explanation of GS. **a** The pharyngovertebral veins and the posterior nasopharyngeal region drainage route. **b** The drainage through the pharyngobasilar fascia and anterior atlanto-occipital membrane to join with the periodontoidal venous plexus and epidural veins

connection of arterial anastomoses [10, 11]. This anatomical architecture provides a hemotogenous route for the transport of peripharyngeal septic exudates to the upper cervical spinal structure which results in hyperemia and laxity of transverse and alar ligaments (Fig. 1). Transverse ligament is the most important and strong structure in the anatomical region of atlanto-axial joint which holds the dens in neutral position by attaching on either side at the lateral masses of atlas (Fig. 2a–b). However, alar ligaments serve also additional support in the stability of this articulation which bilaterally originates from the lateral side of odontoid to the anterolateral part of the foramen magnum (Fig. 3) [12, 13].

In this retrospective analysis, we present 16 pediatric patients, to the best of our knowledge, as the largest series in the English literature with Grisel's syndrome. The goal of this study is to present the diagnostic examinations, simple treatment modalities, and prognosis of these patients with the anatomical mechanism of this rare syndrome.

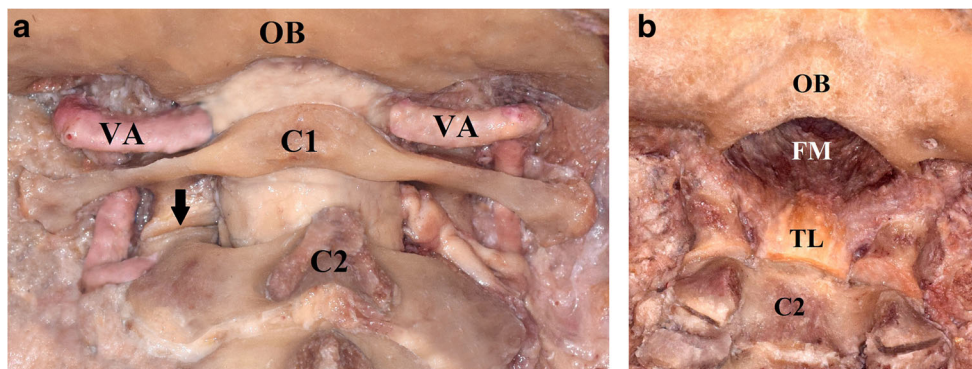


Fig. 2 Posterior view of the craniovertebral junction has been dissected on cadaver. **a** Atlantoaxial joint has been shown on the left side. OB, occipital bone; VA, vertebral artery; C1, posterior arcus of the Atlas, C2, posterior arcus of the axis; arrow indicates atlantoaxial joint. **b** The

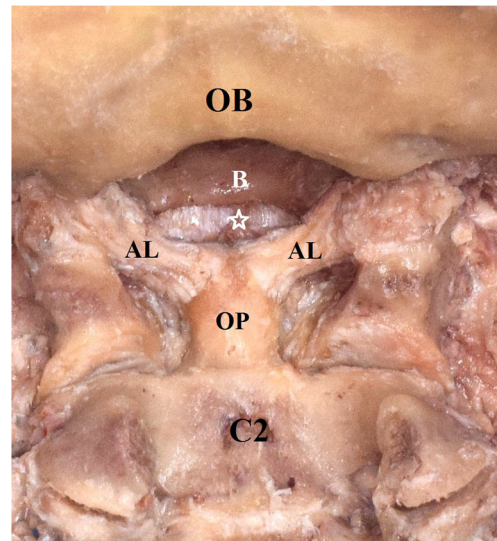


Fig. 3 The view of occipitocervical region was seen for showing the alar ligaments. OB, occipital bone; AL, alar ligament; OP, odontoid process; B, basion, C2, axis; ★, apical ligament

Materials and methods

During a 10-year period (May 2008–May 2018), 16 pediatric patients with Grisel's syndrome were admitted to the Department of Neurosurgery and Otorhinolaryngology, University of Mersin, Faculty of Medicine, Turkey. Clinical data of these patients were reviewed retrospectively and analyzed along with the diagnosis, treatment modality, and prognosis. The clinical parameters of all patients are shown in Table 1.

In addition to clinical cases, five cadavers were dissected for demonstrating atlantoaxial region and parapharyngeal area in a step-wise manner. Anatomical dissections were performed with a Leica, Wild M695 surgical microscope.

panoramic view of the atlantoaxial region has been demonstrated for showing the relationship between the odontoid process and transverse ligament. Note the thickness of the transverse ligament. OB, occipital bone; FM, foramen magnum; TL, transverse ligament; C2, axis

Table 1 The clinical parameters of GS patients

| No | Age, sex | Etiology | Duration | Diagnosis | Type | Treatment |
|----|----------|---------------|----------|-----------|------|--------------------------------|
| 1 | 6, boy | URTI | 2 months | CT | I | SAR + P + SA + I ^a |
| 2 | 7, girl | URTI | 1 day | CT | II | SAR + P + SA+I ^a |
| 3 | 14, boy | URTI | 1 day | CT | II | SAR + P + SA + I ^a |
| 4 | 9, girl | URTI | 7 days | CT | III | SAR + P + SA + I ^a |
| 5 | 9, boy | URTI | 6 days | MRI | II | SAR + P + SA + I ^a |
| 6 | 10, girl | Adenoidectomy | 5 days | MRI | II | SAR + P + SA + I ^a |
| 7 | 7, girl | URTI | 5 days | MRI | I | AR + P + SA + I ^b |
| 8 | 6, girl | URTI | 5 days | CT | II | AR + P + SA + I ^b |
| 9 | 14, boy | URTI | 7 days | CT | I | AR + P + SA + I ^b |
| 10 | 10, boy | Tonsillectomy | 7 days | CT | I | AR + P + SA + I ^b |
| 11 | 11, girl | Tonsillectomy | 6 days | CT | I | AR + P + SA + I ^b |
| 12 | 7, boy | URTI | 2 days | CT | I | AR + P + SA + I ^b |
| 13 | 9, boy | URTI | 1 day | MRI | I | AR + P + SA + I ^b |
| 14 | 3, boy | URTI | 5 days | MRI | II | SAR + SC + SA + I ^c |
| 15 | 13, girl | URTI | 3 days | CT | I | AnR + P + SA + I ^d |
| 16 | 9, boy | URTI | 4 days | MRI | I | AR + P + CA + I ^e |

^a SAR + P + SA + I: Reduction under sedoanalgesia + Philadelphia collar + Sulbactam/Ampicillin + Ibuprofen
^b AR + P + SA + I: Reduction under analgesia + Philadelphia collar + Sulbactam/Ampicillin + Ibuprofen
^c SAR + SC + SA + I: Reduction under sedoanalgesia + soft collar + Sulbactam/Ampicillin + Ibuprofen
^d AnR + P + SA + I: Reduction under anesthesia + Philadelphia collar + Sulbactam/Ampicillin + Ibuprofen
^e AR + P + CA + I: Reduction under sedoanalgesia + Philadelphia collar + Clindamycin/Ampicillin + Ibuprofen

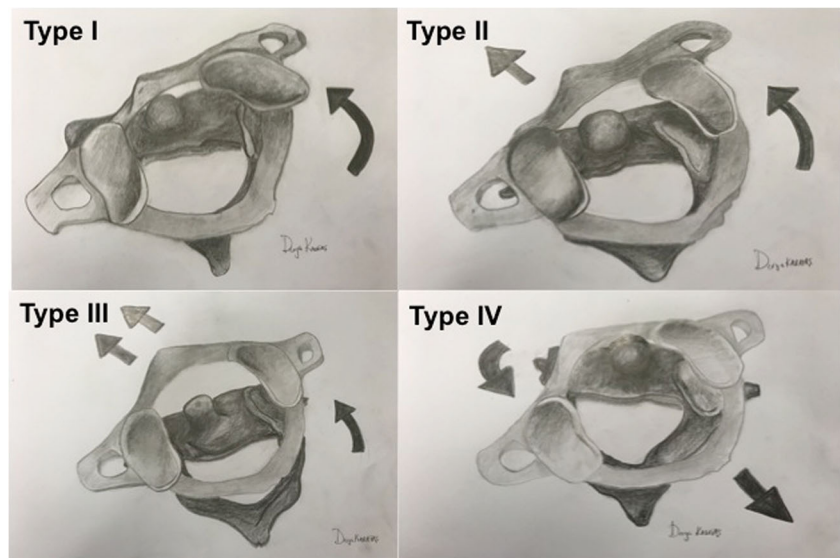
Results

There were nine boys and seven girls GS patients among 16 children. The mean age was 9.0 with the range of 3–14. URTI was the most common etiological factor in our series with 13 patients (81.25%), followed by three patients (18.75%) with surgical interventions (2 tonsillectomy, 1 adenoidectomy). All patients were presented with painful torticollis as the initial

symptom without any neurological deficit in the first week of admission, except one.

All of the cases were evaluated in terms of the laboratory analysis (complete blood count, sedimentation rate, and C-reactive protein) according to clinical diagnosis. Then, the radiological investigation was done with X-ray of cervical spine, CT, or MRI. The Fielding and Hawkins classification for atlantoaxial instability in GS was evaluated by CT or MRI

Fig. 4 The artistic illustration presents Fielding-Hawkins classification. Type I: The atlas is rotated on the odontoid process without anterior displacement. Type II: The atlas is rotated with 3–5 mm anterior displacement. Type III: Atlas rotation with anterior displacement greater than 5 mm. Type IV: Rotatory fixation with posterior displacement of C1



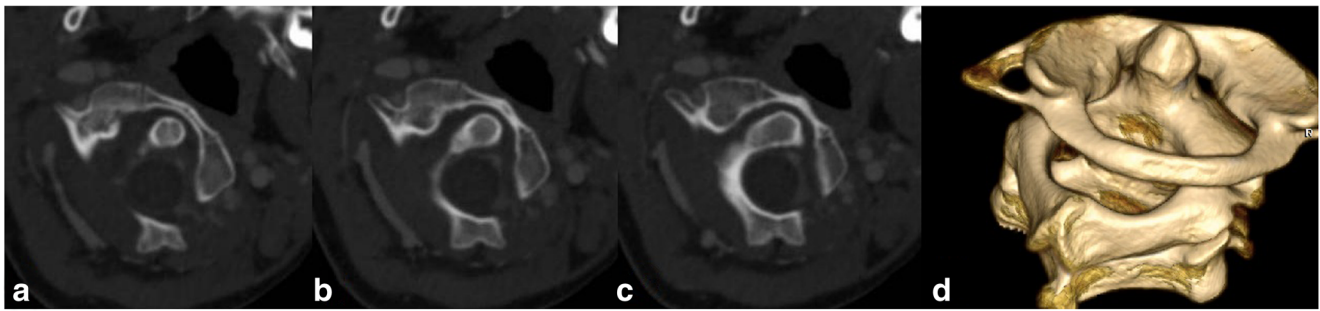


Fig. 5 **a–c** The consecutively axial CT scan showing rotatory atlantoaxial subluxation without anterior displacement of Atlas (Fielding-Hawkins type I subluxation). **d** The demonstration of type I subluxation with 3D reconstruction of CT. Note the severity of rotatory atlantoaxial subluxation

[14, 15]. This classification was illustrated by drawing in Fig. 4. Nine patients were type I (Fig. 5), six were type II (Fig. 6), and only one was type III (Fig. 7) atlantoaxial rotatory subluxation (AARS) according to Fielding and Hawkins classification.

The primary treatment for GS consisted of antibiotic therapy (Sulbactam/Ampicillin 50 mg/kg/day, Clindamycin 30 mg/kg/day) except one patient because of adverse effect, anti-inflammatory therapy (Ibuprofen 5–10 mg/kg/day), bed rest, and external fixation for 2–4 weeks after reduction under analgesia, sedoanalgesia, or general anesthesia. Initial laboratory examinations showed elevated values for CRP and leucocytes with a normalization during the first week of antibiotic therapy.

The reduction was achieved in eight patients under analgesia and in seven patients under sedoanalgesia. Only one patient with accompanying retropharyngeal abscess was reduced under general anesthesia following the otorhinolaryngological surgery.

Discussion

GS is a rare syndrome in pediatric patients as a complication mainly seen after URTI and various otolaryngological

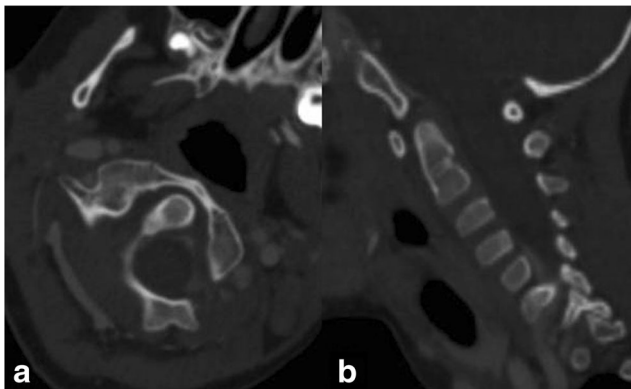


Fig. 6 **a** The axial CT image demonstrates rotatory atlantoaxial subluxation (Fielding-Hawkins type II). **b** Anterior atlantodental interval is measured between 3 and 5 mm on the sagittal view of CT

interventions including, adenoidectomy, tonsillectomy, and also mastoidectomy. There is no clear evidence for the pathogenesis of GS especially in children among the published series. The pathogenesis of GS is explained in two different hypothesis by Battiatà et al. [16]. The first hypothesis is the alar and transverse ligament laxity seen in pediatric age group. According to the second hypothesis, the transport of the inflammatory agents to cervical muscles and atlantoaxial joint via the pharyngovertebral plexus is easier in pediatric age groups than adults. As a result, it causes cervical muscle spasm and subsequent subluxation. In addition, large sized skull, partial development of cervical muscles and uncinat process, shallow and horizontal perspective of facet joints, and also excess amount of lymph nodes are the explanations to identify the reasons of GS especially for children under 4 years [4, 5, 17]. In our series, only one patient (6.25%) under 4 years of age exist. Gender predominance was not found among the patients. Deichmueller and Welkoborsky reported 67% of cases after surgery and 33% URTI in series of 12 patients of GS [1]. Meanwhile, Subach et al. reported a series of 63.6% of GS patients after URTI and 36.4% after adenotonsillectomy [18]. Mezue et al. mentioned all 13 GS patients occurred after URTI in their series [19]. Hence, the etiological factors show differences from series to series in the published studies. In our series, 13 patients (81.25%) after URTI and 3 patients (18.75%) after surgery (2 tonsillectomy, 1 adenoidectomy) were diagnosed as GS.

The usual presenting symptoms of GS are cervical pain with head tilt and limited and painful neck movement. In the first week after infection or surgery, the painful torticollis is the leading symptom of this syndrome similar to the literature. In our experience, almost all of the patients were admitted within the first week with painful torticollis, except one [3, 20]. According to clinical data, painful torticollis in the following days of surgery or infections could be also the gold standard for diagnosis of GS. Despite of this fact, the presence of AARS is verified to be a necessity for radiological investigation. The anterior atlantodental interval is 2–3 mm in children physiologically. If this interval exceeds more than 5 mm in children, the atlantoaxial instability should be considered [1, 21]. The atlantoaxial subluxation can exactly be shown

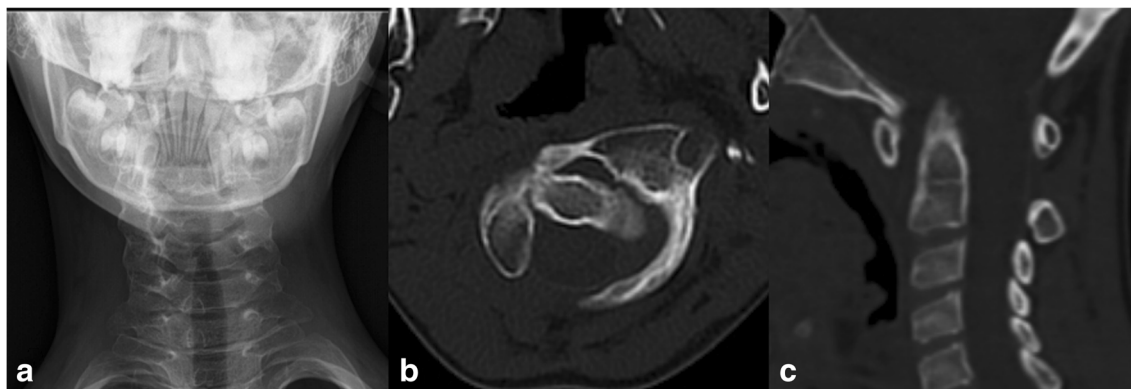


Fig. 7 **a** Anteroposterior X-ray shows “cock-robin” position. **b** Axial CT indicates rotatory atlantoaxial subluxation without anterior displacement of Atlas (Fielding-Hawkins type III subluxation). **c** Anterior atlantodental interval is measured more than 5 mm on the sagittal view of CT

only in CT scan especially with 3D images. In the radiological evaluation of GS, MRI could also be used which has advantages for demonstrating soft tissues. On the other hand, hyperemia of transverse or alar ligament after infection seen in GS can mimic retropharyngeal space abscesses around this region which may lead to unnecessary surgeries [1]. In our series, the final diagnosis of GS was achieved in ten patients with CT, in six patients with MRI. If GS can be diagnosed without any doubt by only considering patient’s history and clinical examination, CT scan is not recommended in children by the authors due to harmful effects of radiation. Therefore, after all of our clinical experience, MRI can safely be preferred instead of CT for children in the diagnosis and follow-up period in the last patients of our series (Fig. 8).

The antibiotic and anti-inflammatory treatments were used in all patients prior to the diagnosis. Analgesia was sufficient to make successful reduction for seven patients, whereas eight patients needed to be given sedative analgesia (Midazolam 0.05–0.1 mg/kg) for reduction. There was only one patient reduced under general anesthesia with parapharyngeal abscess whom had undergone surgery by otorhinolaryngologist. In the literature, Pilge et al. reported a new technique of reduction with manual reposition under general anesthesia in five patients with good clinical results. (1 type I, 2 type II, 2 type III) [20]. However, according to our study, general

anesthesia is not necessary for reduction in early diagnosed pediatric patients independent of GS types. Analgesia or sedoanalgesia is adequate for successful reduction. After reduction, one patient under 4 years was followed with soft neck collar. Philadelphia collar was used for 2–4 weeks in the rest of the patients for external fixation. Finally, the treatment algorithm consist of reduction and external fixation for early diagnosed GS children by Philadelphia collar under analgesia or sedoanalgesia together with antibiotic and anti-phylogistic treatment for effective therapy. Neither neurological deficits nor deformities were seen in our series.

Conclusion

Painful torticollis following URTI or surgery could be a gold standard for diagnosis of GS in pediatric population. Although CT is standard in detection of AARS, MRI should be the initial radiological evaluation especially in children due to harmful effects of radiation. Meanwhile, early diagnosis and appropriate treatment is vital in the management of GS. The conservative management including anti-inflammatory and anti-phylogistic treatment with reduction may be suggested as the choice of therapy, if initiated early, without the necessity of invasive procedures like Halo and deformity surgeries.

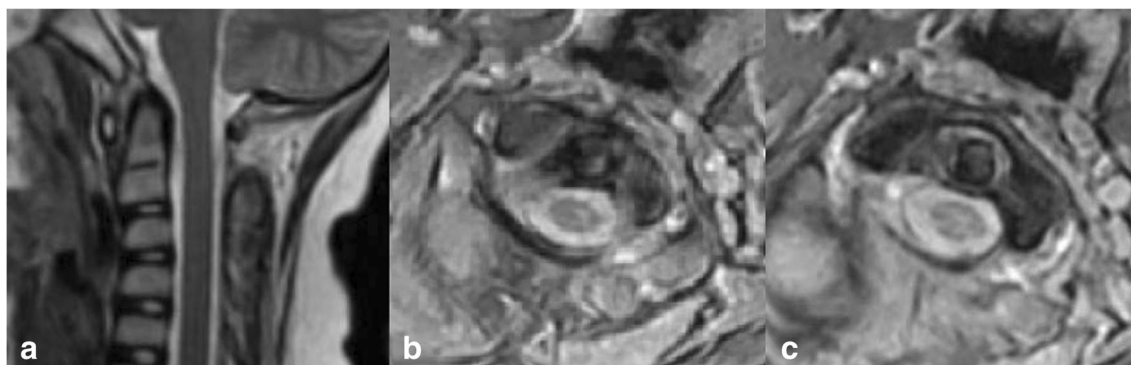


Fig. 8 The sagittal (**a**) and axial (**b**, **c**) T2 MRI shows rotatory atlantoaxial subluxation (Fielding-Hawkins type I)

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