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Case Report

The Importance of Early Diagnosis and Appropriate Treatment in Grisel's Syndrome: Report of Two Cases

Grisel Sendromunda Erken Teşhis ve Uygun Tedavinin Önemi: İki Olgu Sunumu

Ahmet DAGTEKIN[1], Engin KARA[2], Yusuf VAYISOGLU[3], Aslihan KOSEOGLU[1], Emel AVCI[1], Derya TALAS[3], Celal BAGDATOGLU[1]

[1]Mersin University, Faculty of Medicine, Department of Neurosurgery, Mersin, Turkey
[2]Mersin University, Faculty of Medicine, Department of Radiology, Mersin, Turkey
[3]Mersin University, Faculty of Medicine, Department of Otorhınolaryngology, Mersin, Turkey

Correspondence address: Ahmet DAGTEKIN / E-mail: dagtekin69@yahoo.com

ABSTRACT

Grisel's syndrome designates subluxation of atlanto-axial joint unrelated to trauma or bone disease. Atlanto-axial subluxation is seen after the upper neck inflammatory processes of head and neck region. Moreover, this rare clinical entity has been observed after various otolaryngological surgical approaches including adenoidectomy, tonsillectomy and mastoidectomy. An 8-year-old girl and 14 year-old boy presented with complaints of painful torticollis. The first patient was operated on at another health center due to adenoid hypertrophy. She was referred to our clinic on the postoperative 5th day with painful torticollis. The diagnosis was established by computerized tomography and three-dimensional reconstructions. The second patient had a history of throat infection. Radiological investigation demonstrated atlanto-axial subluxation. The patients were treated with external fixation, antibiotic therapy, muscle relaxants, and anti-inflammatory therapy. Early diagnosis and appropriate treatment resulted in a good outcome in our patients. Grisel's syndrome must be considered in the differential diagnosis in children with painful torticollis associated with upper respiratory tract infection and after various otolaryngological surgical procedures. We report two cases of Grisel's syndrome which emphasize on the importance of early diagnosis for appropriate and successful treatment.

KEYWORDS: Grisel's syndrome, Non-traumatic atlanto-axial subluxation, Torticollis, Computerized tomography

ÖZ

Atlantoaksiyal eklemin travma ya da kemik patolojileri gibi nedenlere bağlı olmayan subluksasyonu Grisel Sendromu olarak adlandırılır. Oldukça nadir görülen bu sendrom boyun bölgesinin enflamatuvar hastalıkları ya da cerrahi girişimleri sonrası ortaya çıkabilir. Başka bir sağlık merkezinde adenoid hipertrofisi nedeniyle cerrahi girişim uygulanan ve postoperatif 2. gün ağrılı tortikolis gelişen sekiz yaşında kız hasta, postoperatif 5. gün yakınmalarının artması nedeniyle kliniğimize sevk edilmiştir. Ağrılı tortikolis yakınması ile kliniğimize başvuran ondört yaşında erkek hastanın hikayesinde ise 1 hafta önce geçirilmiş boğaz enfeksiyonu vardı. Radyolojik incelemeler sonucu her iki hastada da atlantoaksiyal subluksasyon tespit edildi. Servikal boyunluk ile eksternal fiksasyon, antibiyotik, kas gevşetici ve antienflamatuar tedavi uygulanan hastaların yakınmalarında hızla düzelme gözlendi. Boyun bölgesinin enflamasyonla seyreden hastalıkları ve cerrahi girişimleri sonrasında gelişen ağrılı tortikolisin ayırıcı tanısında Grisel Sendromu akılda tutulmalıdır. Bu iki olgu nedeniyle non-travmatik atlantoaksiyal subluksasyonun erken tanı ve uygun tedavisinin önemi vurgulanmıştır.

ANAHTAR SÖZCÜKLER: Grisel sendromu, Non-travmatik atlantoaksiyal subluksasyon, Tortikolis, Bilgisayarlı tomografi

INTRODUCTION

Spontaneous subluxation of the atlanto-axial joint following naso-pharyngeal inflammation is known as Grisel's syndrome (1,5,7,10,17). The syndrome was first described by Bell (4). He reported a case of death from atlanto-axial subluxation which resulted in spinal cord compression in a patient with a syphilitic ulceration of the pharynx (1). Grisel's syndrome is generally associated with head and neck infection such as pharyngitis, adenotonsillitis tonsillar abscess, cervical abscess and otitis media (1,3). It has also been observed after numerous otolaryngological procedures such as tonsillectomy, adenoidectomy and mastoidectomy (1,11).

This syndrome is most frequently seen in children (1,3,4). The most widely accepted pathogenesis for Grisel's syndrome is the inflammation that causes hyperemia which has been attributed to the laxity of the transverse and alar ligaments (15). The usual presentation of this clinical entity is with cervical pain, head tilt, limited and painful neck movement.

The diagnosis is made by clinical findings and radiological investigations.

CASE REPORTS

Case 1: An 8-year-old girl was referred to our clinic with painful torticollis. She was operated in another health center due to adenoid hypertrophy. On the postoperative second day, she had received muscle relaxants and anti-inflammatory therapy for painful neck movement. She was referred to our clinic on the postoperative 5th day for painful torticollis. Neurological examination was normal. X-ray of the cervical spine and CT were performed for diagnosis. Atlanto-axial rotary was suspected by plain roentgenograms of the cervical spine (Figure 1). CT scan of the upper cervical spine revealed atlanto-axial rotary subluxation (Figure 2), confirmed by 3-D reconstruction (Figure 3, 4). A reduction of subluxation was performed under analgesia. The patient was placed in a cervical collar and received a 2-week course of i.v. antibiotic therapy (sulbactam + ampicillin 50 mg/kg/day). External cervical fixation was extended to 4 weeks. At the end of the first week, the patient's painful torticollis resolved completely. Atlanto-axial rotational subluxation was reduced on the CT examination at the end of the 4th week (Figure 5).

Case 2: A previously healthy 14-year-old boy was admitted to our clinic with painful torticollis. He had no history of trauma, but had received antibiotic therapy for throat infection for one week. Neurological examination revealed no abnormalities. Antero-posterior radiographs of the cervical spine show the typical head tilt in this patient which is known as the cock-robin position (Figure 6). Cervical spine axial CT showed atlanto-axial subluxation (Figure 7). Antibiotic therapy (sulbactam + ampicillin 50 mg/kg/day iv), muscle relaxants, and anti-inflammatory therapy were started and a cervical collar was applied. The painful torticollis had resolved and axial CT examinations showed no abnormalities at the end of the second week in the follow-up examination (Figure 8).

Figure 1: Antero-posterior radiographs of the cervical spine show the typical head tilt (cock-robin position) in the first case.

DISCUSSION

Non-traumatic subluxation of the atlanto-axial joint is an unusual and poorly understood status which occurs mainly in children (1). The main stabilizer of the atlanto-axial joint is

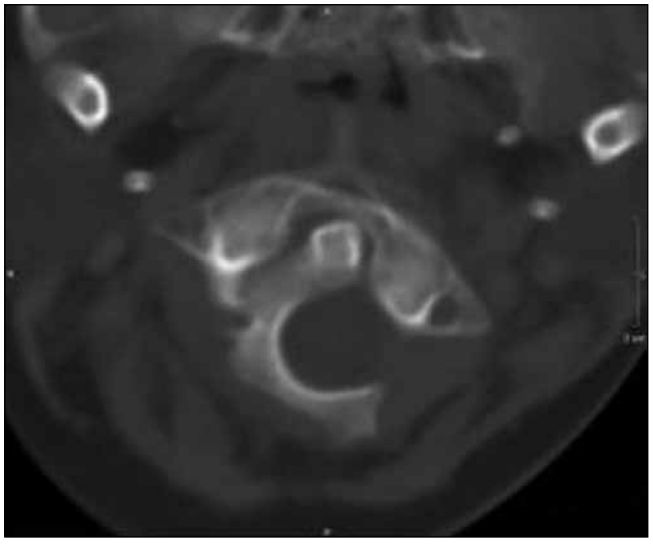

Figure 2: Axial CT scan showing rotatory atlanto-axial subluxation with anterior displacement of atlas (Fielding Type II subluxation).

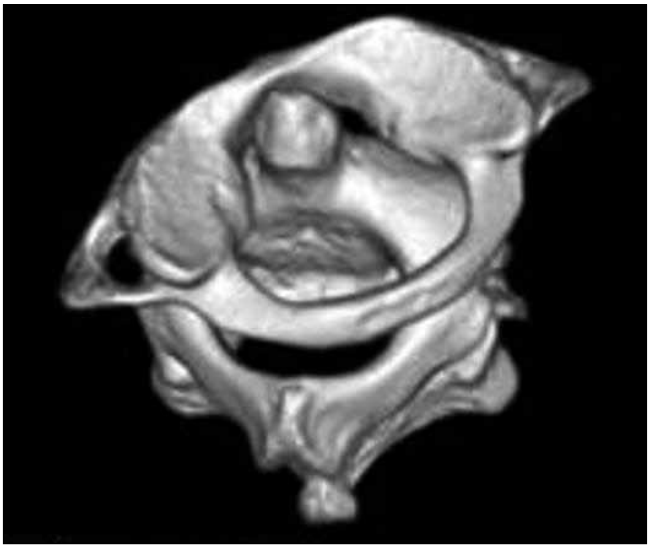

Figure 3: Three-dimensional reconstruction CT scan showing rotatory atlanto-axial subluxation.

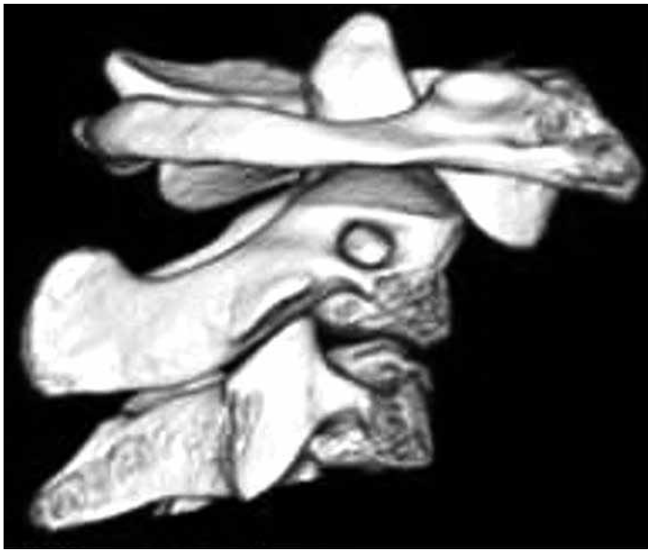

Figure 4: Three-dimensional reconstruction CT scan showing anterior displacement of atlas.

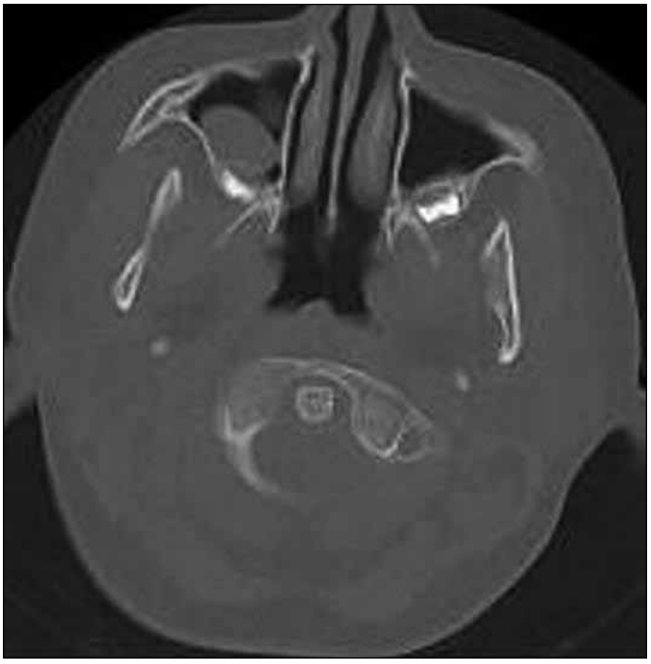

Figure 5: Axial CT scan showing the recovery of atlanto-axial subluxation.

Figure 6: Antero-posterior radiographs of the cervical spine show the typical head tilt (cock-robin position) in the second case.

Figure 7: Axial CT scan showing rotatory atlanto-axial subluxation (Fielding Type I subluxation).

the transverse ligament and the other stabilizers are the alar ligaments (17). The probable pathogenesis causing laxity of above mentioned ligaments leading to subluxation of the atlanto-axial joint is explained with several phenomena. Some of these factors including larger head size, weaker cervical muscles, looser ligament and joints, shallower and more horizontally placed facet joints and existence of a lymphatic system with a greater number of retropharyngeal lymph nodes also help to explain the reason of dominance of this disease in children, especially under the age of 4 (3,4).

Pharyngo-vertebral veins drain to the periodontoidal venous plexuses. Due to the direct connection between the periodontoid venous plexuses and pharyngo-vertebral veins, a hematogenous pathway may be provided for the transport of peripharyngeal septic effusions to the atlanto-axial ligaments.

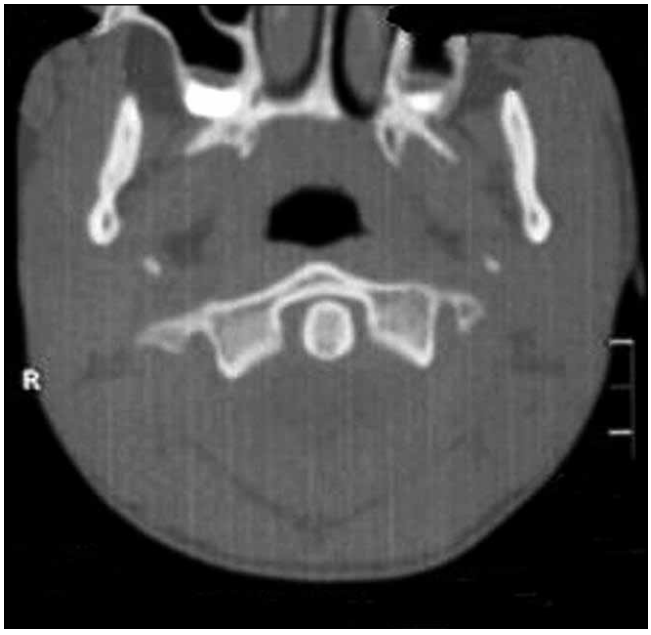

Figure 8: Axial CT scan showing the recovery of atlanto-axial subluxation.

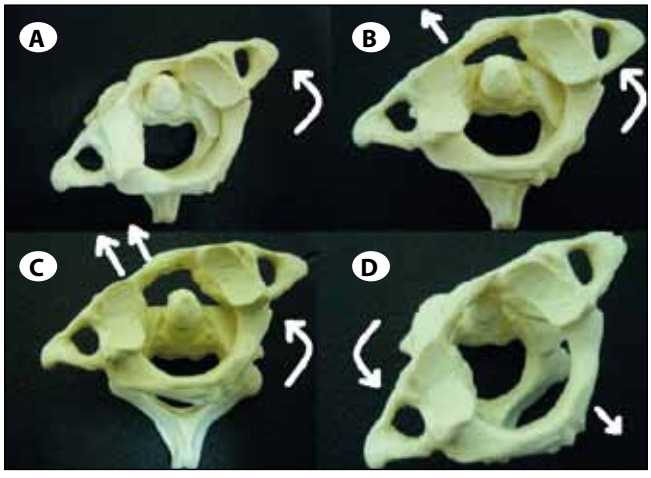

Figure 9: Fielding classification of atlanto-axial rotatory subluxation. **A:** Type I, the atlas is rotated on the odontoid without anterior displacement, **B:** Type II, the atlas is rotated with 3-5 mm. anterior displacement, **C:** Type III, rotation of the atlas with anterior displacement greater than 5 mm., **D:** Type IV, rotatory fixation with posterior displacement of atlas.

Inflammation leading to hyperemia, generally associated with head and neck infections or surgical procedures, results in a laxity of the transverse and alar ligaments (15).

Fielding has classified four different types of non-traumatic rotatory atlanto-axial subluxation. In Type I, the atlas is rotated on the odontoid without anterior displacement. In Type II, the atlas is rotated on one articular process with 3-5 mm. anterior displacement. Type III consists of rotation of the atlas with anterior displacement greater than 5 mm. Type IV is explained by rotatory fixation with posterior displacement of atlas (1,4,16) (Figure 9A-D). Type I and II subluxations are most commonly seen and have no neurological deficits. Type III and IV may be associated with spinal cord compression and serious neurological deterioration. Our first case presented with Type II subluxation (Figure 2) and the second patient was consistent with Type I subluxation (Figure 7).

Grisel's syndrome usually manifests with torticollis, cervical pain, head tilt and restricted-painful neck movements (9,10). Torticollis may occur shortly after the onset of the head and neck infection or it may follow otolaryngological procedures (1,6,14). Painful torticollis is noticed on the postoperative second day in our first case. In the second case, torticollis was reported to begin on the seventh day of the throat infection.

Early diagnosis of Grisel's syndrome is of utmost importance due to the neurological complications, if the disease is overlooked and/or treated inappropriately or late. Radiological investigations are very valuable for the management of atlanto-axial subluxation. These evaluations are plain radiographs, CT and magnetic resonance imaging (MRI). Plain radiographs can be difficult to interpret but some findings are suggestive (7). Lateral plain radiographs of the upper cervical spine may show an increase in the atlanto-odontoid distance. The normal atlanto-odontoid distance is ≤ 3 mm in adults, and ≤ 5 mm in children (2). CT is the most helpful method in establishing the diagnosis of atlanto-axial subluxation. The best visualization of this pathological state is by 3-D CT (8). MRI is a complementary radiological tool. MRI may also reveal the abnormality of soft tissue and neural structures. Atlanto-odontoid distance was measured as 4 mm in our first case in CT examination. This measurement was in normal range in this age group. However 3-D CT reconstruction revealed Type II non-traumatic rotatory atlanto-axial subluxation. Atlanto-axial rotary dislocation was noticed in the axial CT for the second case.

The primary treatment of early detected Grisel's syndrome is conservative. Conservative treatment includes bed rest, external fixation, antibiotic therapy, muscle relaxants, and anti-inflammatory therapy. If the diagnosis is established, atlanto-axial subluxation must be reducted as soon as possible. The length of time until reduction, has been directly related to the failure of medical treatment and to an increased risk of recurrence or permanent neck deformity (3,12). Delayed diagnosis and treatment may result in painful and permanent neck deformity that may even require surgical therapy (13). In our first case, the subluxation of atlanto-axial joint was thought to be due to previous adenoid surgery. A reduction of subluxation was performed under analgesia. External immobilization was applied to maintain stability of atlanto-axial joint. In the second case, it was thought to be due to the inflammation of the throat infection and external immobilization was performed. Intravenous antibiotic therapy was begun for both patients. Recovery was observed within one week for both of the cases. Early diagnosis and appropriate treatment probably lead to the early recovery in our patients.

Grisel's syndrome is an unusual condition. It must be considered in children with painful torticollis associated with upper respiratory tract infection and after various otolaryngological surgical procedures. We want to emphasize the importance of early diagnosis and treatment of Grisel's syndrome. Diagnosis was performed by clinical findings and radiological investigation. When the diagnosis is established, treatment should be started immediately. Antibiotic therapy and cervical collar are the first choice of treatment.

In conclusion, early diagnosis permitting conservative treatment prevent permanent neck deformity, serious neurological deficit, and extensive surgical approach.

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