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Case Report

Infection-related atlantoaxial subluxation in two adults: Grisel syndrome or not?

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Summary

Grisel's syndrome involves the subluxation of the atlanto-axial joint from inflammatory ligamantous laxity following an infectious process in the head or neck. It is a rare disease usually affecting children, but infrequent adult cases do occur. Today, due to the widened use of antibiotics and availability of MR imaging, some cases presenting with neck pain, resulting from the infectious inflammation of C1 and C2 but without pronounced subluxation, can be a challenging problem for most neurosurgeons. Several theories have been proposed to explain the pathogenesis of inflammatory subluxation. The primary treatment of Grisel's syndrome is medical. The underlying infectious organism must be isolated and appropriate antibiotics must be prescribed. The subluxation is reduced in holter or skeletal traction. This paper reports two cases of infection-related atlanto-axial subluxation in two adults. The literature on this subject is briefly reviewed.

Keywords: Atlantoaxial subluxation; Grisel's syndrome; infection; respiratory tract.

Introduction

Atlanto-axial instability in association with pharyngeal infection, which has come to be known as Grisel's syndrome was described earlier by Bell. It may occur in association with any condition that results in hyperaemia and relaxation of the transverse ligament of the atlanto-axial joint [3]. Several common otolaryngial entities have been associated with the syndrome: pharyngitis, nasopharyngitis, adenotonsillitis, tonsillar abscess, parotitis, cervical abscess, and otitis media. Several theories have been proposed to explain the pathogenesis of inflammatory subluxation. Witteck thought that a metastatic inflammatory effusion caused ligamentous stretching and subluxation. Grisel, however, suggested that subluxation occurred on the basis of muscle spasm. The mechanism for Grisel syndrome is mostly believed to be hyperaemia following infections or surgical trauma that leads to decalcification of the anterior arch of the atlas and laxity of the anterior transverse ligament between the atlas and the axis. Anatomical studies have demonstrated the existence of a peri-odontoidal vascular plexus which drains the posterior superior pharyngeal region. No lymph nodes are present in this plexus, so septic exudates may be freely transferred from the pharynx to the C1–C2 [2, 3, 4]. Most patients complain of neck stiffness and pain on attempted motion. Dysphagia is also a frequent complaint. Patients usually are neurologically intact. The diagnosis is made by clinical examination and radiological findings. Early diagnosis is very important for the success of the treatment [1, 3, 5]. This study aims at contributing to the relevant literature with two adult cases of infection related atlanto-axial dislocation, to briefly discuss the pathogenesis, the chance of differential diagnosis of Grisel like syndromes, and the kind of treatment modalities of Grisel's syndrome which is very unusual in adults.

Case report

Case 1

A previously healthy 31-year-old woman presented with persistent neck pain. One week before admission, she had a sore throat and high fever. There was no history of trauma. Physical examination revealed a mild right torticollis while throat examination was normal. There was no neurological impairment. The white blood cell count was 7.600 with a normal differential, and the erythrocyte sedimentation rate was at 10 mm/h. Recurrent throat cultures showed

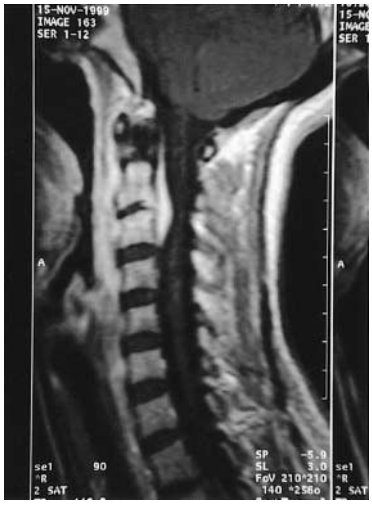

Fig. 1. Sagital T1 contrast enhanced MRI shows a mass infiltrating the anterior C1-2-3 subligamentous region

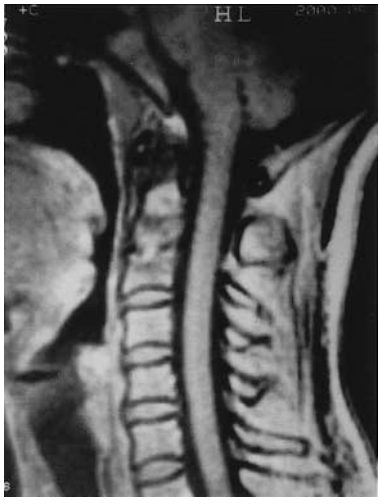

Fig. 2. Sagital T1 contrast enhanced MRI shows disappearance of the mass after treatment

normal flora. All immunological markers were normal. The plain cervical x-rays and computed tomography (CT) showed normal bone structure. Magnetic resonance imaging (MRI) revealed a mass infiltrating the anterior C1-2-3 subligamentous region (Fig. 1). Despite the absence of atlanto-axial subluxation, due to the clinical correlation, the patient was thought to have Grisel's syndrome in progress. The patient received a 6-week-course of wide spectrum antibiotics and was stabilised in a Philadelphia collar. Six weeks later, the patient's neck pain and stiffness completely resolved, and no mass was detected on MRI (Fig. 2).

Case 2

A 77-year-old woman was admitted to our clinic with a complaint of persistent neck pain, which had been ongoing for one year. There was a history of lung tuberculosis 30 years before her admission. She

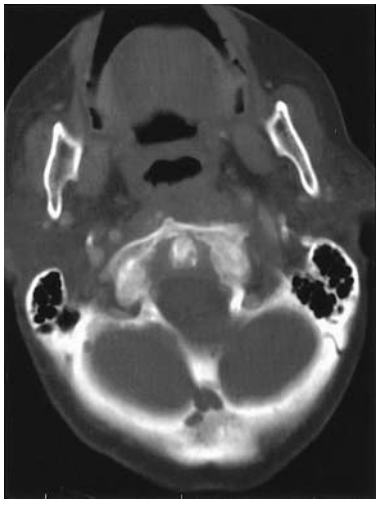

Fig. 3. CT of the neck demonstrating atlanto-axial subluxation and erosion of the odontoid process

denied any history of injury, trauma or infection. Physical and neurological examinations were normal. The white blood cell count was 4.100, while erythrocyte sedimentation rate was 8 mm/h. All immunological markers were normal. The plain cervical x-rays and CT revealed subluxation of C1 on C2 with erosion of the odontoid process (Fig. 3). MRI showed an anterior prevertebral mass on gadolinium enhanced sections. A transoral biopsy was planned to confirm the diagnosis and to rule out the possibilities of the tuberculoma or malignant infiltration. Pathological examination showed a non-specific inflammatory reaction associated with Grisel's syndrome. The patient received a 6-week-course of wide spectrum antibiotics and was stabilised in a Philadelphia collar. Six weeks later all her symptoms had resolved, and on CT, atlanto-axial interval reduced but rotational subluxation did not recover. The patient refused to have any surgical or non-surgical treatment, and is currently being followed-up.

Discussion

Although Grisel's syndrome was first described in 1830s, there has been long-standing and unresolved controversy regarding the pathogenesis, diagnosis, and the best treatment modalities of the syndrome. The primary stabiliser of the atlanto-axial joint is the transverse ligament, attached to the lateral posterior portion of the anterior arch of the atlas, forming the posterior support of the odontoid process, preventing excessive shifts of C1 on C2. The paired alar ligaments are secondary stabilisers preventing excessive rotation [5]. Several theories have been proposed to explain the pathogenesis of the syndrome. The theories of metastatic inflammation and muscular spasm are not accepted by most authors today. The mechanism for Grisel's syndrome is generally believed to be hyperaemia following infection or surgical trauma that leads

to decalcification of the anterior arch of the atlas and laxity of the anterior transverse ligament. Recent anatomical studies supporting this theory demonstrate the existence of a peri-odontoidal vascular plexus which drains the posterior superior pharyngeal region and having no lymph nodes, so septic exudates may be freely tranverse from the pharynx to the C1-2 articulation [4]. This rare disease usually affects children, but is infrequent in adults. Patients generally seek treatment for progressive unrelenting throat and neck pain followed by torticollis. Neurological complications occur in approximately 15% of cases [1, 2, 3, 4, 5]. Especially when the clinical presentation is not obvious, i.e. when there is no history of sore throat with high fever, no Sudeck's sign (the spinous process of C2 is palpable away from the midline at the site opposite the dislocation), and no cock–robin sign (torticollis with the chin turned away from the subluxated side, the head is tilted towards the affected side and flexed anteriorly making it difficult to open the mouth), the early diagnosis is very difficult. In such cases if the patients are not followed up, they will be at high risk of neurological complications and increased atlanto-axial subluxation. The important point in the early diagnosis is that clinical presentation and findings should be evaluated carefully and the possibility of the Grisel's syndrome should be taken into consideration. Radiological evaluations are postero-anterior and lateral cervical spine films with an open mouth view, CT, and MRI. CT and MRI should be performed with contrast to reveal a mass or swelling. On the lateral view, there may be an increase of the atlantodental interval, the space between the posterior margin of the anterior arch of atlas, and the odontoid process. On plain films normal evaluation does not rule out subluxation. CT is useful in defining bony destruction and misalignment. MRI which shows a soft tissue in this region, may hint at the possibility of the existence of Grisel's syndrome. In both cases of this study, soft tissue swelling was demonstrated. Radiological investigations are not diagnostic especially in the early period of the disease and recurrent investigations or dynamic studies may be required. Active and passive movements of the neck can be very painful and may cause neurological complications during dynamic studies. Fielding has characterised four different types of rotatory atlanto-axial subluxation. The atlas is rotated on the odontoid, and there is no anterior displacement of the atlas in type 1. In type 2, the atlas is rotated on one lateral articular process with 3 to 5 mm of anterior atlas displacement. Type 3 consists of rotation of the atlas with anterior displacement greater than 5 mm. Type 4 is characterised by rotation and posterior displacement of the atlas [4]. Our first case has no subluxation while the second case is classified as Fielding type 1. Another issue which we faced with our first case was that the patient had all clinical findings of Grisel's syndrome, and no evidence of atlanto-axial subluxation, making it difficult to decide whether we should diagnose the patient as a Grisel's syndrome case. Based on the clinical and MRI findings, we diagnosed the patient as having Grisel's syndrome. With proper antibiotic treatment, the mass disappeared and clinical findings resolved. Early diagnosis allowed early treatment and cure. The widespread use of MRI studies make an early diagnosis of the cases suffering from neck pain and presenting atypical Grisel Syndrome like symptoms possible. Grisel Syndrome, in its classical form, was defined two centuries ago for the first time, and it was very difficult to achieve an early diagnosis of the syndrome at those times. No matter what it should be named, Grisel syndrome or not, clinical presentation due to the infection involved in this syndrome must be borne in mind for early diagnosis and an effective treatment modality. Today, in order to define the aetiology of this syndrome, looking for entities described in the earlier descriptions of the disease may not be fully helpful due to the reasons mentioned above. In addition, the use of wide-spectrum antibiotics in upper respiratory tract infections and the use of highly effective antibiotics in the early periods of soft tissue infections, the infections invading C1-2 areas are suppressed for a long time. Thus, the transverse ligament involvement and subluxation can be prevented in the early stages. In the second case, however, because of the unclear clinical findings, Grisel's syndrome was not considered as a diagnosis at first, but in order to rule out tuberculosis or malignant infiltrations, transoral biopsy was performed and as a result the diagnosis was established as Grisel's syndrome. Transoral biopsy provides a firm and early diagnosis in such cases. Treatment should be individualised depending on the degree and duration of the subluxation as well as the early diagnosis. In addition to antimicrobial therapy, the dislocation should be reduced. If the anterior transverse ligament is intact, cervical traction with a hard cervical collar for 6 to 8 weeks is usually sufficient. If the ligament is deficient, cervical skeletal traction with Gardner-Wells tongs attached to the skull may be required to effect

reduction. Immobilisation of the cervical spine Minerva cast or halo vest is recommended to allow the ligaments to heal. If patients have subluxation lasting longer than 3 months, primary C1-2 fusion is recommended to decrease any neurological sequelae [1, 2, 3, 4, 5].

Conclusion

According to our limited experience and in the light of the literature on the diagnosis of the syndrome:

1. Patients presenting with neck pain and torticollis following respiratory tract infection should be studied considering the possibility of Grisel's syndrome even if they are adults.
2. The atlanto-axial subluxation may not constitute a rule in early-diagnosed cases if clinical presentation relates to Grisel's syndrome.
3. Early diagnosis is the most important issue in the treatment and prevents serious neurological sequelae.

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Comments

The authors report 2 cases of inflammatory pathology of the atlanto-axial vertebral complex. In the first case neck pain and mild torticollis developed 1 week after a throat infection. I am not sure that I would have ordered an MRI scan after such a short history. The findings of a posterior longitudinal thickening and enhancement from C1 to C3 support the author's contention that an early inflammatory process was present, as does the resolution of the findings following neck immobilization. The second case is an elderly lady with chronic neck pain due to atlanto-axial subluxation with a prevertebral mass. In this case, transoral biopsy yielded non-specific inflammatory tissue.

The authors have described an interesting entity, the pathophysiology of which is speculative. While atlanto-axial subluxations are known to occur in the pediatric age following oropharyngeal infections, an increasing number of cases are being described in adults. The review of the subject is therefore timely.

Z. H. Rappaport

This paper reports two cases of Grisel's syndrome – atlanto-axial subluxation due to ligamentous laxity following a local infectious process in the head or neck. This rare condition was found in two adults, a 31 year old woman and a 77 year old man, and although diagnosis was only evidenced in the latter by transoral biopsy indirect evidence is presented and the case reports with history, clinical findings and image diagnostics are well presented. Also the presentation of the syndrome and the discussion is sufficient and relevant, but I miss in the discussion clear indications on when to consider performing MRI in patients with some of the signs of the syndrome. At the same time focusing the readers attention to the syndrome is very relevant and recommended for publication.

K. Dons

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