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Case Report

Grisel's syndrome in adults: A case report

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ABSTRACT

Background: The non traumatic, post inflammatory atlantoaxial rotatory instability, also known as Grisel's syndrome is a relatively rare condition usually affecting children. Adult cases are rare and even less frequently reported with separate case reports describing a single patient. Although antibiotic treatment and close neurological monitoring seem to be the gold standard of care, there is no general consensus on the optimal timing and extent of the surgical treatment.

Case Description: We present a case of C1-C2 spondylitis, secondary to retropharyngeal abscess, without atlantoaxial instability on initial evaluation that progressed to C1-C2 subluxation with rapidly developing myelopathy 3 months after optimal antibiotic therapy and complete clinical and biochemical remission.

Conclusion: Grisel's syndrome is a rare condition in adults with secondary instability in spite of successful antibacterial treatment, which requires decompression and delayed surgical fixation in our case.

Keywords: Atlantoaxial rotatory instability, Atlantoaxial subluxation, Grisel's syndrome, Surgical treatment

INTRODUCTION

Post inflammatory C1-C2 subluxation was described in the first half of the 19th century by Charles Bell it was later named after Grisel who published a paper on two patients with retropharyngeal inflammatory involvement of the C1 anterior arch, C2 dens, and ligamentous complex.[5] It is a rare condition affecting children and adolescents usually related to the upper respiratory tract infections, tonsillitis, surgery for adenoidectomy, palatoplasty, and a few other pathological processes.[4] Pathoanatomical findings suggest that an existing vascular bridge between the periodontoidal plexus and the retropharyngeal drainage veins is the main channel for the metastatic dissemination of infectious emboli.[9] The subsequent clinical development is thoroughly investigated in pediatrics and anatomical peculiarities such as more horizontal and shallow facet joints, incomplete bone mineralization, and innate ligamentous laxity are characteristics for this group being cited as the main predisposing factor for the development of non traumatic atlantoaxial rotatory instability (AARI). There are three theories behind the pathogenesis of this entity: first, granulation developing secondary to metastatic infectious effusion, with the growing granulomatous tissue stretching the ligamentous complex and displacing the bony structures; and second, irritation causing muscle spasms leading to asymmetrical subluxation; at last, post infectious laxity of the transverse ligament coupled with decalcification of the C1 arch and lysis of the ligament insertion points which are widely

accepted to be the main pathogenic mechanisms.[10] Usual clinical presentation in adults includes, but is not limited to: cervical pain/tenderness, torticollis, and Sudeck's sign, with as many as 15% of reported cases progress with neurological deficits.[1,2,8]

CASE DESCRIPTION

A 48-year-old male was treated at our institution between April and September 2018, no history of intravenous drug use or HIV infection was noted. He was referred to the department of neurosurgery with a 1 month history of the upper cervical region pain radiating toward the shoulders and proximal brachia, tenderness, and increased rigidity of the neck musculature. The patient reported a history of oropharyngeal bacterial infection that was interpreted as strep throat by the general practitioner, for which the patient received a 10-day course of antibiotic. Although local inflammation was resolved the patient continued to feel under the weather, complaining of night sweats general lack of energy and later on – neck pain and tenderness that was previously described. After a cervical magnetic resonance imaging (MRI) was performed [Figure 1] by a consulting neurologist the patient was referred to our clinic for further investigation and treatment.

Initial complete blood count (CBC) was significant for leukocytosis – 24.12 × 10^9/L, ESR was 70 mm/h, and C-RP was 92.10 mg/L. In light of the lab test results and the history of infectious pharyngitis, we postulated that the bacterial infection was incompletely resolved and has now spread to the retropharyngeal space and the C1-C2 complex. Blood cultures were negative and broad-spectrum antibiotic treatment was ineffective. During the diagnostic work-up, standard dynamic X-rays were obtained to ascertain the presence of instability and the need for surgical C1-C2 fixation [Figure 2a]. No evidence of hypermobility in the segment was evident, so we decided to proceed with rigid external cervical orthosis and antibiotic treatment according to microbiological findings. We decided to perform a transoral biopsy of the retropharyngeal mass to isolate the pathogenic organism – Staphylococcus aureus. After a 3-week course of vancomycin, 1 g twice and ceftriaxone 2 g once daily i.v. the patient showed significant clinical improvement. CBC, ESR, and C-RP were all within referent range. The patient was then discharged and ordered to continue an antibiotic treatment of gentamycin 160 mg daily i.m. for another 3 weeks as an outpatient, with weekly CBC, ESR, and C-RP. He was advised to wear the rigid cervical collar for another 3 months after which he was scheduled for a control visit.

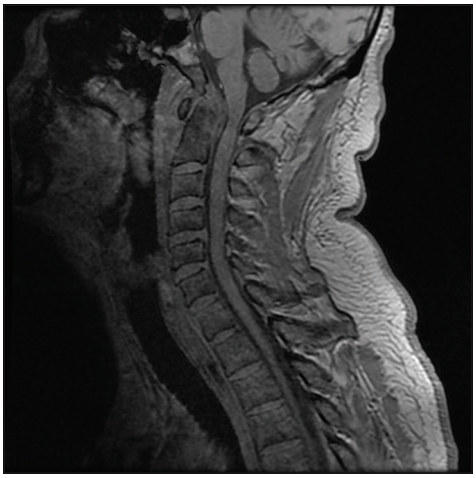

Figure 1: Sagittal T1 magnetic resonance imaging – evidence of a retropharyngeal mass expanding to the C1-C2 complex, yet the C1 anterior arch – C2 dens distance is preserved. The dens are centered without evidence of instability.

However, 2 weeks before the appointment the patient became symptomatic for the upper cervical cord compression – increasing pain in the neck, occiput and shoulders, weakness in both arms, gait disturbances, and rigidity and hyperreflexia for both legs. A new emergency MRI was ordered that showed complete resolution of the retropharyngeal abscess and the C2 spondylitis; however, signs of spinal cord compression with myelopathy as well as instability were evident with atlantodental interval (ADI) out of range [Figure 2b], later confirmed with X-rays (not shown). The native CT of the occipitocervical junction afterward showed significant ectopic calcification of the transverse and alar ligaments and subsequent pseudoarthrosis [Figure 2c].

At this point, we decided to perform an emergent C1 posterior decompression with C1 lateral mass and C2 isthmus screws fixation and fusion [Figure 3a and b].[6] The patient went on to make complete neurological recovery. On the 3-month follow-up, his examination was uneventful and the patient has no subjective symptoms.

DISCUSSION

Although extremely rare in adults Grisel's syndrome should always be suspected when there is clinical evidence of paravertebral infectious processes, with secondary C1-C2 involvement. Even without evidence of instability on initial assessment these patients can deteriorate rapidly because of secondary atlantoaxial rotational hypermobility. Initial imaging studies that suggest soft-tissue inflammation, but no atlantoaxial hypermobility or subluxation, are not definitive and these patients need close follow-up. A standard lateral X-ray of the cervical spine could be sufficient to evaluate progressing instability. In our case a patient with no significant C1-C2 subluxation went on to develop Fielding Grade III AARI in <3 months.[3] One of the main challenges, we faced when managing this case was that we had a patient with all clinical and imaging prerequisites for Grisel's syndrome without any AARI

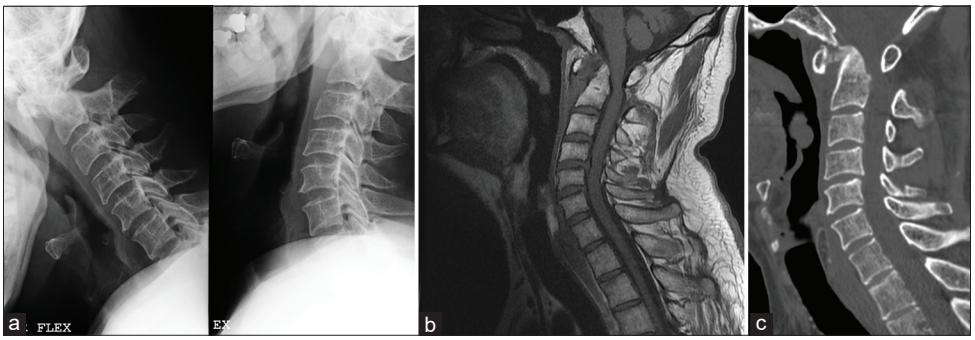

Figure 2: (a) The atlantoaxial complex is stable on lateral dynamic X-rays with atlantodental interval (ADI) in range (<3 mm). (b) Sagittal T1 magnetic resonance imaging – no retropharyngeal mass persist, but the C2 dens is significantly dorsally displaced causing central cervical canal stenosis. ADI is pathologically increased (14 mm) with evidence of subluxation and C1-C2 stenosis. (c) Sagittal computed tomography (bone) – evidence of alar and transverse ligament involvement with calcification of the osteoligamentous complex.

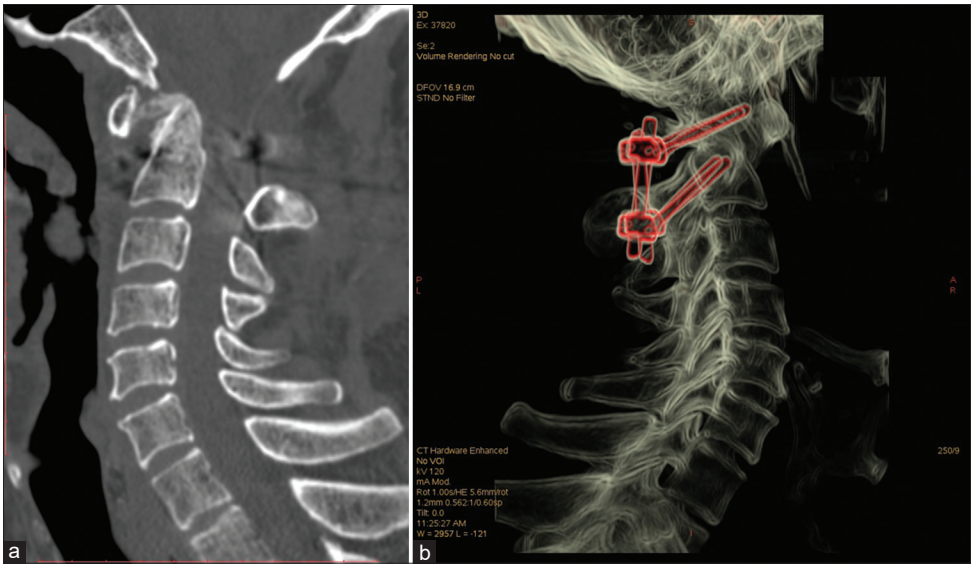

Figure 3: (a) Corresponding postoperative sagittal computed tomography (CT) (bone) – adequate decompression of the cervical spinal canal in comparison to the preoperative scans. (b) Postoperative CT reconstruction – C1 lateral mass screws and C2 isthmus/pedicle instrumentation.

on initial examination, an issue documented by other authors as well.[7] This makes extensive and expensive surgical procedures hard to rationalize and further investigation is required to determine the rational of early and aggressive surgical management against the conservative "wait and see" approach.

CONCLUSION

Patients presenting with cervical vertebral syndrome with a history of upper respiratory tract infections should be suspected for Grisel's syndrome, irrespective of their age. Lack of AARI on initial examination should not exclude the diagnosis and it might progress to instability with spinal compression once confirmed. There is not enough clinical evidence to support early, aggressive surgical management, although our experience and other case reports suggest that surgical immobilization of the C1-C2 complex may produce better short-term results and prevent later dynamic instability.

Declaration of patient consent

Patient's consent not required as patient's identity is not disclosed or compromised.

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Conflicts of interest

There are no conflicts of interest.

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