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Case Report

COVID-19 causing Grisel's syndrome? A rare case report

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ABSTRACT

Background: Grisel's syndrome was initially described by Grisel in 1930 to explain the case of a non-traumatic atalanta-axial subluxation (C1 over C2). Exact aetiopathogenesis is unclear with the patient usually presenting as a painful acquired torticollis.

Clinical Description: A 9-year-old male child presented with restriction of neck movements, cough for 2 months and tongue deviation for a week. The patient had increased deep tendon reflexes in bilateral upper and lower limbs with bilateral exaggerated plantar reflexes. Power in bilateral lower limbs was 5/5. The patient also had a history of COVID-19 infection 3 weeks back which was managed conservatively.

Management: X-ray of the cervical spine revealed subluxation of the atlanto axial joint with an increased atlanto dens interval of 7.5 mm. Magnetic resonance imaging of the Cervical region showed an increased atlanto dens interval of 7.6mm. The patient was immobilised immediately with a hard cervical collar. The patient was started on Inj. Ceftriaxone 850 mg IV twice daily for 21 days, followed by another 21 days of Tab. Cefixime 100 mg twice daily. A repeat X-ray of the cervical spine showed a reduction in atlanto axial dens interval and an improvement in the patient's condition.

Conclusion: Although the diagnosis of Grisel's syndrome could be challenging at times, it is imperative to manage patients with rare diseases swiftly and precisely to prevent permanent future disabilities. Whether COVID-19 was the trigger for the development of Grisel's syndrome is up for debate and potential area of new research.

Keywords: COVID-19 complication, Atraumatic Atlanto-axial dislocation, Paediatric age group

INTRODUCTION

Grisel syndrome was first described by a French otolaryngologist, Grisel in 1930 to explain the association between inflammation and atraumatic atlantoaxial subluxation. Since then it has become a well-documented but rare clinical entity and is being defined as 'a non-traumatic, fixed rotatory subluxation of C1 on C2 (atlantoaxial).'[1] The aetiology and pathogenesis remain unclear with no universally accepted mechanism for the subluxation. The following theories are put forth:[2]

1. Watson and Jones suggested that it was due to the resorption of the attachment of the transverse ligament to the atlas by an inflammatory cause. The reported case had the reappearance of a decalcified anterior arch of the atlas after the reduction and treatment of the infection.
2. Wittek's take on Grisel syndrome involved experiments on a rabbit's knee where there was laxity of the ligaments when pyarthrosis (inflammatory zone) was induced by injection of turpentine solution and thus theorised that such environment would cause distention of the

ligaments in the cervical region leading ultimately to the subluxation.

3. Finally, a theory put forth by Parke involved both the above mentioned theories. He described that the pharyngovertebral veins anastomose with the lymphatics of the retropharyngeal space, thus explaining the possibility of an upper respiratory infection causing Atlanto Axial subluxation.
4. Battiata and Pazos recently published in 2004 that the reason behind the increased incidence of Grisel in the paediatric age group involves two hits: (i) Pre-existing cervical ligamentous laxity (most specifically-the transverse ligament) and hypermobility of C1 on C2 (evidenced by increased atlas-dens interval of 4.5 mm in children versus 2.5 mm to 3 mm in adults), (ii) Parke's theory of pathogenesis by pharyngo-vertebral veins thereby explaining the muscle spasms and subluxation.

In this study, we describe a case of Grisel's Syndrome with a possible aetiology of COVID-19, due to its presentation during the 2nd peak of COVID-19 wave in our country.

CASE REPORT

A 9-year-old male child presented to our Outpatient Department on 29 November 2021 with complaints of cough, fever and painful restriction of neck movements for 2 months. On further inquiry, the cough was insidious in onset, associated with purulent sputum production, mild quantity, non-blood tinged, associated with fever, moderate grade, intermittent type which aggravated on exposure to cold weather and relieved on taking Tab. Paracetamol 500 mg SOS. However, there was no significant history of weight loss, nor an evening rise in temperature. A striking history of restriction of neck movements was brought out by the parents, which was also insidious in onset, aggravated for 5 days with travel and no history of trauma. There was a deviation of tongue to the left, for 5 days, acute in onset [Figure 1]. On questioning further, it was revealed that there was no vision abnormality, smell disturbances, facial asymmetry, bowel or bladder disturbances, hearing disturbances or any history of frequent falls. The child could also feel warmth over his face while taking a bath. There was also no history of difficulty in swallowing, indicating that the cranial nerves I to XI are not likely affected. On 4 October 2021, the patient tested and reported to have elevated antibodies for COVID-19 indicating past infection (with the father testing positive for COVID-19 at the same period in the past) and at a later date, the patient had complaints of chronic cough as mentioned above. Previous hospital visits lead to the diagnosis of the patient with Multisystem Inflammatory Syndrome in Children who was treated with Tab. Prednisolone, Tab. Azithromycin, Tab. Cefuroxime and Tab. Aspirin orally. The patient was symptomatically feeling better initially. The symptoms reappeared 2–3 weeks after the discharge. Later, they approached our hospital for further management. The patient was then admitted to our hospital and upon extensive clinical examination, increased deep tendon reflexes on all four limbs were seen. The knee jerk reflex was 3+ bilaterally, along with increased bilateral plantar reflexes. The sensory system examination was normal. No other focal neurological deficits were seen other than the deviation of the tongue to the left.

Management and outcome

In routine blood investigations, the total WBC Count was 15,300/mL. Suspecting pulmonary TB, a chest X-ray PA was done which was normal. Following which sputum sample for ZN (Ziehl-Neelsen staining) Staining and GeneXpert-*Mycobacterium tuberculosis* Bacilli was sent and was reported to be negative. Antineutrophil cytoplasmic antibodies panel was also sent in view of vasculitis, due to the outside High-Resolution Computed Tomography report suspecting Wegener's Granulomatosis, and was reported to be negative. Automated culture and sensitivity of blood were sought in view of infection and were also negative. For further diagnoses of the chest pathology, a bronchoalveolar lavage was needed which could not be done due to restriction of neck movements. The patient was thus started on Inj. Ceftriaxone 850 mg twice daily to cover both Gram-positive and negative strains of bacteria. A Neurosurgery opinion was obtained in view of the restriction of neck movements and was diagnosed as Grisel's Syndrome. The patient was immediately immobilised with a Philadelphia cervical collar. The cough and fever spikes reduced. Following a decrease in symptoms, magnetic resonance imaging (MRI) of head-and-neck was done and reported to have increased Atlanto-dens. MRI of the cervical spine was

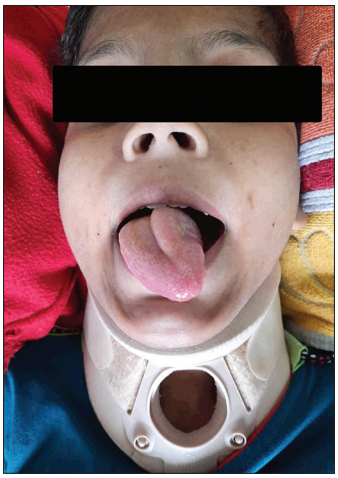

Figure 1: Deviation of tongue to left.

performed using Philips Achieva 1.5 Tesla MR scanner and the following pulse sequences were acquired non-contrast: Sagittal T1W, T2W, STIR and axial T2W, STIR and DWI. MRI showed loss of normal lordotic curve of the cervical spine with cortical irregularity noted involving the left occipital condyle and a posterior arch and superior articular surface of the atlas. It is associated with anterolateral subluxation of the Atlanto-occipital joint with Atlanto dens interval measuring 7.6 mm(increased)-[Figure 2]. Cervical lymphadenopathy was also present.

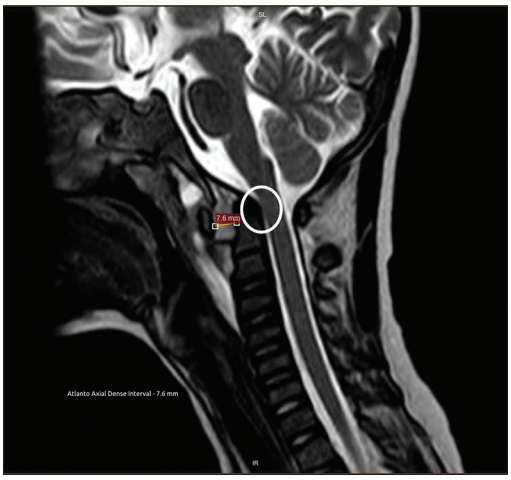

Figure 2: T2W MRI of Cervico-Vertebral Junction depicting increased Atlantodens interval of 7.6 mm.

Neurosurgeons advised surgery if worsening of the current deviation or the appearance of new focal neurological deficits. The patient was continuously monitored for 21 days during the IV antibiotic administration. Following which the patient was discharged at the request and was advised to start on Tab. Cefexime 100mg orally in the morning and night for the next 21 days, and continue neck immobilisation as before. After 6 weeks of antibiotics, the patient no longer had neck pain, and there was an improvement in tongue deviation. Subsequent Cervico Vertebral Junction X-rays showed a decrease in atlanto dens interval [Figure 3]. The patient is currently feeling better and was asked to follow up after 3 months.

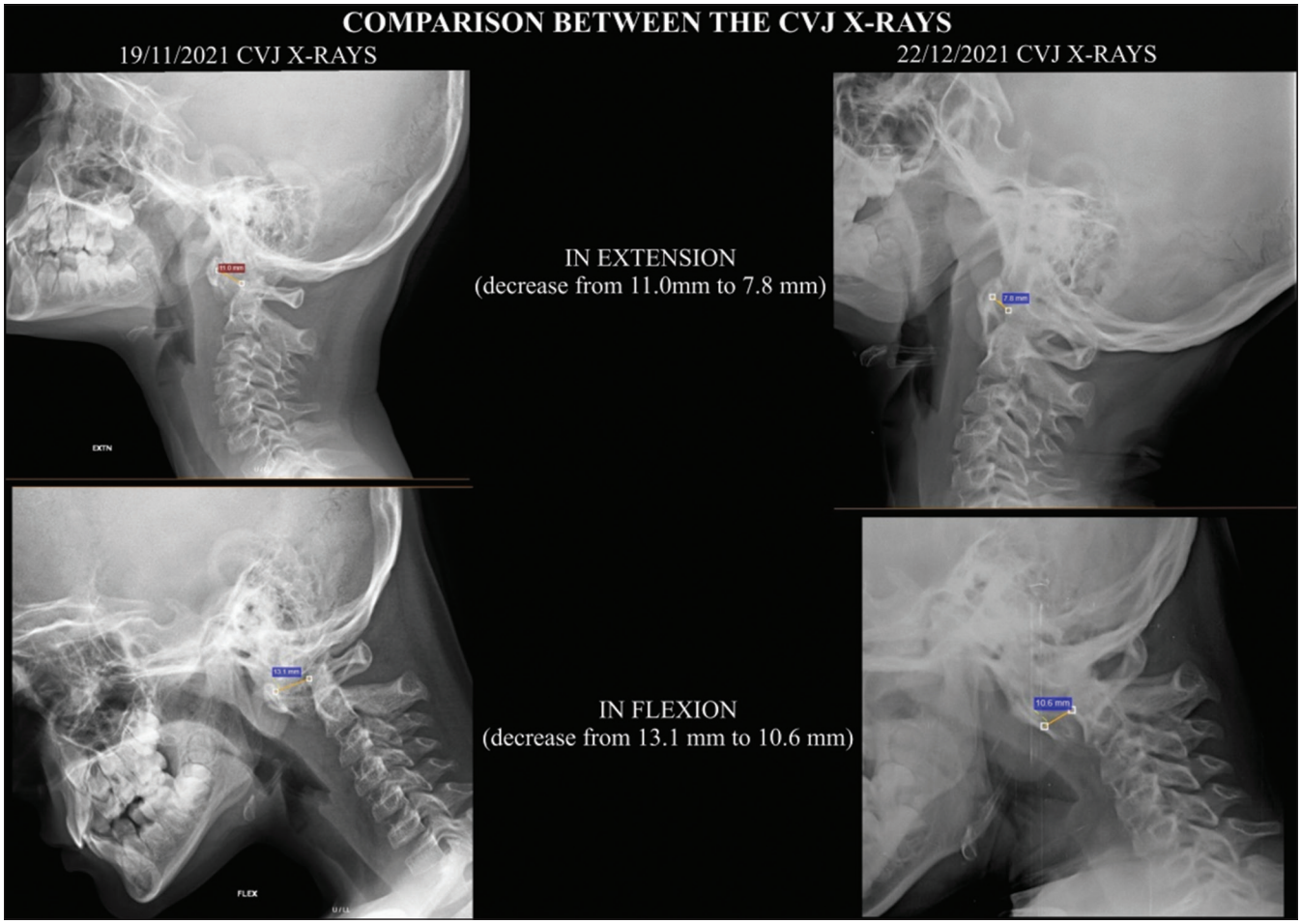

Figure 3: Comparison of X-ray images of Cervico Vertebral Junction.

DISCUSSION

Atraumatic subluxation of C1 over C2 is defined as Grisel's syndrome. Although the aetiology and pathogenesis seem unclear, up to 68% of the patients are under the age of 12 and 90% are under the age of 21 with no gender predominance. It is most commonly secondary to a post-operative complication (26.9%) or an upper respiratory infection (18%). The patients of Grisel's syndrome typically present with a painful restriction of neck movements (torticollis), and other non-specific signs of infection like fever. On examination, a positive Sudeck's sign (a palpable spinous process of the axis on the side opposite the dislocation) is observed commonly. Neurological signs are seen in 15% of the cases.[3]

Management of Grisel's depends on the classification by Fielding,[3] usually with contrast CT of the cervical region to help distinguish it from an abscess. MRI of the head and neck is also done to evaluate any focal neuro deficits and to act as a baseline for spinal involvement.

Gourin believed that the initial treatment of Grisel's syndrome is usually managed medically with the administration of IV antibiotics.[4] However, Fielding and Hawkings classified the rotatory subluxation consists of four types, and proposed their concurrent treatment options as given below: [3]

- Type 1 – Rotatory fixation with no anterior displacement with odontoid acting as the pivot point – traction and immobilisation with soft collar
- Type 2 – Rotatory fixation with no anterior displacement of 3–5 mm, one lateral articular process acting as a pivot – traction and immobilisation with a hard collar
- Type 3 – Rotatory fixation with anterior displacement of more than 5 mm – surgical halo fixation
- Type 4 – Rotatory fixation with posterior displacement – open fixation.

However, the treatment options differ when neurologic symptoms are present, with most of the studies suggesting immediate open fixation for the same.

The prognosis for Grisel syndrome depends on many factors:

1. The lower success rate for patients with AAS for more than 3 weeks in duration
2. Progression is seen when reported late
3. Better prognosis when treated early with intravenous antibiotics and sedatives.

CONCLUSION

The etiology for Grisel's Syndrome in our patient was not clear, it ultimately left us to ponder upon the possibility of COVID19 being the one. Whether COVID19 is one of the new etiology of Grisel's syndrome is up for debate and warrants new research, with some researchers concluding that it is considered as one.[5]

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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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