



Case report

Conservative approach for treatment of Grisel's syndrome after resection of lymphadenitis tuberculosis of the neck: A rare case report

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ARTICLE INFO

Keywords:

Grisel's syndrome
Acquired torticollis
Conservative
Minerva cast

ABSTRACT

Introduction: Grisel's syndrome is a rare condition characterized by nontraumatic rotatory subluxation of the atlantoaxial joint, which was caused by previous inflammation around the head and neck. It is usually seen in children and signed as torticollis. There was no consensus for management, yet early diagnosis and treatment is paramount.

Presentation of case: A 5-year-old girl came to outpatient clinic complaining of wry neck 1 day after surgical excision of her TB lymphadenitis and got worsen by time. There was no history of trauma around the neck. Examination under general anesthesia and CT scan revealed acquired severe torticollis consistent with Grisel's Syndrome Fielding type 3 accompanied by TB lymphadenitis, and TB myositis of the neck. Manipulation under general anesthesia and immobilization using Minerva cast followed by Lerman Cervicothoracic Orthosis was conducted.

Discussion: This patient was diagnosed with Grisel's syndrome and underwent conservative treatment consisting of reduction under general anesthesia and immobilization using Minerva cast for 6 weeks. The patient was then applied Lherman Cervical Thoracic Orthosis (CTO) halo brace for another 3 months. Anti-tuberculous drug was given to control tuberculous infection. Eight months follow-up showed neither residual deformity, neck pain, nor movement limitation of the neck.

Conclusion: Grisel's syndrome has excellent result that is treated with conservative treatment using reduction under general anesthesia and Minerva cast.

1. Introduction

Grisel's syndrome is a rare syndrome characterized by nontraumatic rotatory subluxation of the atlantoaxial joint, which is usually seen in children. Patient typically presents with acquired torticollis [1,2]. Distension and irregular laxity of the atlanto-axial articulation ligaments result from the direct spread of inflammation from the pharynx and nasopharynx or after ear, nose, and throat (ENT) surgery, head and neck infections. It may contribute to dysfunction of the cervical spine with catastrophic neurological sequelae, including death [2–4].

Grisel's syndrome is a rare complication of head and neck disease [5]. Moreover, this syndrome may lead to poor prognosis if left untreated. Early disease entity identification is critical to determining the best treatment for the patient. The goal of Grisel's syndrome treatment is to correct deformity while preserving neck motion. This syndrome has been treated conservatively and surgically with release and cervical

fusion [6,7]. Nevertheless, due to its rare incidence there is as yet a guideline how to best treat this condition. Unsuitable and irrational treatments can cause morbidity and even death.

In this paper, we present a case of Grisel's syndrome which has been successfully treated with conservative approach. This study aims on elucidating the outcome of this treatment with 8 months follow-up. This study was performed in line with SCARE checklist guideline [8].

2. Presentation of cases

In our outpatient clinic, a 5-year-old girl had wry neck a month after a biopsy for a neck lump. She was a healthy child who had been ill for 9 months with night sweats, mild fever, loss of appetite, and nausea. A negative Mantoux test and urinary infection were ruled out. She got B12 coenzyme and cyproheptadine. The patient gained weight well after treatment. However, 5 months after starting therapy, she developed

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<https://doi.org/10.1016/j.ijscr.2021.106452>

Received 9 August 2021; Received in revised form 22 September 2021; Accepted 22 September 2021

Available online 1 October 2021

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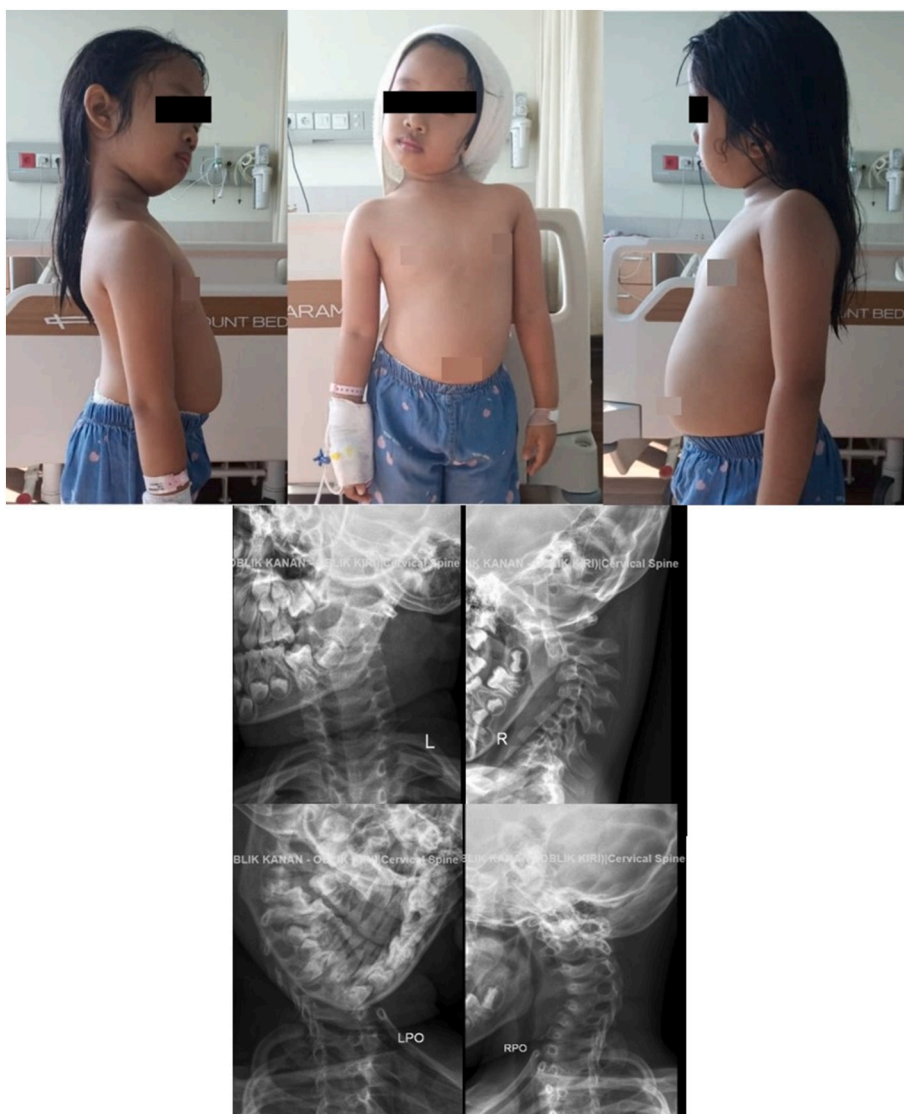


Fig. 1. Clinical picture and plain neck radiograph when the patient first visited our spine center.

several neck lumps that grew from 12 mm to 22 mm in just one month. After a negative interferon gamma release assay (IGRA), the patient underwent excisional biopsy.

Histopathological result revealed that specific granulomatous lymphadenitis with Dutia langhans cells consistent with tuberculosis infection. She was treated with anti-tuberculosis drugs, then performed surgical excision of the lymphadenopathies. A day after the treatment, the patient complained of headache, the head began to tilt left-sided, fever, nausea, and hoarseness. A brain MRI was ordered which revealed no sign of meningitis. A month after biopsy, the tilting of the neck became worse and the patient underwent laser puncture several times to no avail. Another ultrasound was ordered which revealed shortened sternocleidomastoid muscle, after which, the patient was referred to us.

Physical examination and plain radiographs (Fig. 1) did not give us any useful information so we ordered a CT scan and performed the exam under anesthesia (EUA). EUA had revealed - deformity withstanding - a flexible neck which full range of motion and the tightness on the concave side of the torticollis. Furthermore, the neck could be re-aligned to normal position (Fig. 2). The flexibility, location of soft tissue tightness, and ability to realign all indicated soft tissue fibrosis due to previous procedure and infection. Subsequently, while still in anesthesia, the patient underwent CT-scan which depicted atlanto-axial subluxation

(Fig. 3).

From the CT scan, we concluded that the patient suffered from acquired severe torticollis consistent with Grisel's Syndrome Fielding type 3 accompanied by tuberculous lymphadenitis, and tuberculous myositis of the neck. After a lengthy discussion with her parents, we elected to perform manipulation under general anesthesia and immobilization using Minerva cast. After 6 weeks, the Minerva cast was changed into with Lherman Cervicothoracic Orthosis (CTO) or pinless halo brace for further 3 months (Fig. 4). Eight months follow-up (Fig. 5) revealed excellent neck posture without any residual deformity, neck pain, or movement limitation of the neck (Fig. 5).

3. Discussion

Following removal of lymph node and infection induced by tuberculosis in the adjacent lymph nodes around the neck, this patient developed atlanto-axial subluxation. Spontaneous subluxation of the atlanto-axial joint following peri-pharyngeal inflammation is known as Grisel's syndrome [9]. This syndrome has been linked to rhinopharyngitis, rheumatic diseases, cervical osteomyelitis, and surgical operations such as adenoidectomy, tonsillectomy, and mastoidectomy [4,10]. Distension and abnormal laxity of the ligaments surrounding the atlanto-axial articulation are due to direct spread of inflammation from



Fig. 2. Physical examination under general anesthesia showed that the neck could be re-aligned to normal position and the tightness was over the concave side of the torticollis (yellow arrow). (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

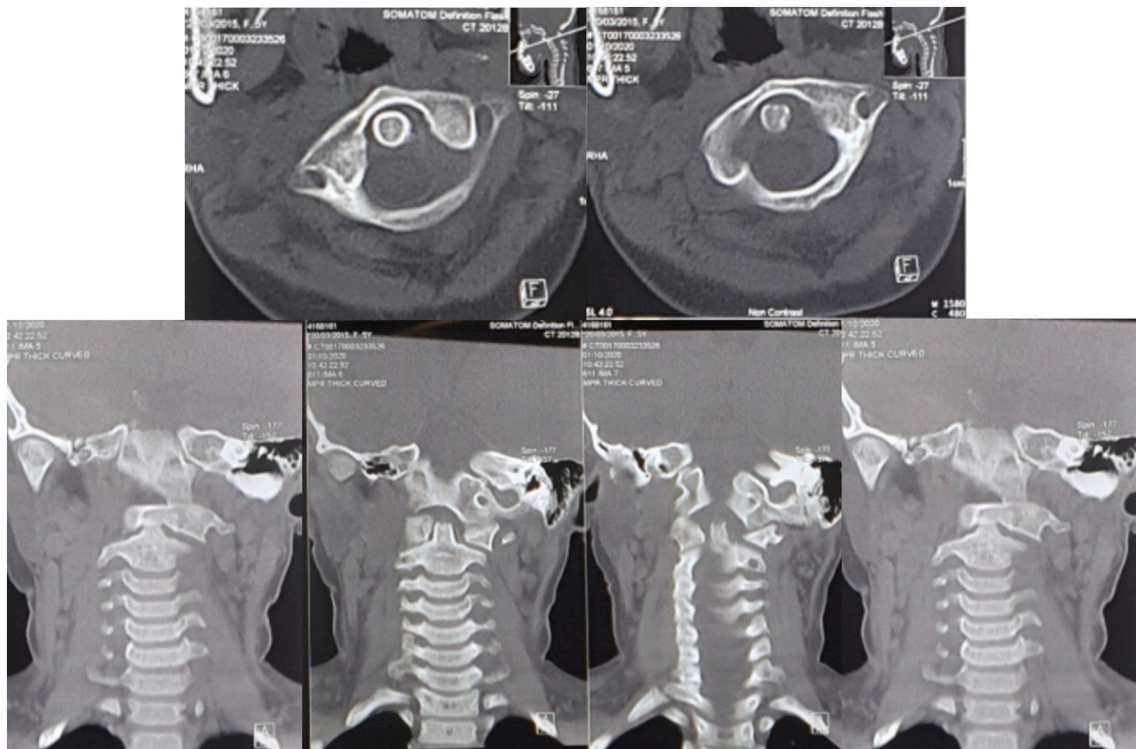


Fig. 3. CT scan of the neck showed atlanto-axial subluxation Fielding type 3.



Fig. 4. A) Minerva cast after manipulation; B) Lherman Cervicothoracic Orthosis (CTO) or pinless halo brace.



Fig. 5. Range of motion after 8 months follow-up.

pharynx and nasopharynx which may result in cervical spine instability with risk of devastating neurological sequelae. Less than 15% of patients with Grisel's syndrome have neurological signs or symptoms, however,

some may present with extreme consequences such as quadriplegia and sudden death.

The basic pathogenesis of Grisel syndrome is pathological elongation

of the ligament around cervical spine joints. This results from inflammatory process or otolaryngological surgery procedure around the head and neck [4]. History of previous neck infection such as tonsillitis, pharyngitis, parotitis, adenoiditis, tonsillar or cervical abscess is the most common cause of inflammation process that may cause Grisel's syndrome [6]. To our finding, resection of lymph node over the neck in combination with ongoing TB lymphadenitis is the most probable cause for this patient.

There are 4 types of atlantoaxial subluxation according to Fielding classification [11]. The first type is the rotation of atlas above axis without presence of anterior displacement. The second type is the rotation of atlas above lateral reticular process with 3–5 mm anterior displacement. The third type is the rotation of atlas with more than 5 mm anterior displacement, and the fourth type is the rotation of atlas with posterior dislocation [6,12].

Grisel's syndrome mostly occurs in childhood because pediatric patients often develop upper airway infection. Children also have bigger head-to-trunk proportion, weaker cervical muscle, more ligamentous laxity, and more horizontally structured of C1-C2 joint. These factors may cause C1-C2 subluxation [13]. The pharyngovertebral veins flow across the posterior pharyngeal wall and nasopharynx to periodontal plexus and atlanto-occipital membrane on both sides. These veins may facilitate a hematogenous septic effusion of a pharyngeal infection to reach the atlantoaxial joint, which further causing subluxation [14].

Our patient had cervical tuberculosis prior to atlantoaxial dislocation. The pathogenesis is unknown. Inflammation and pathology of the ligament in the atlantoaxial joint may cause it. An inflammation of the neck could cause atlas decalcification and anterior transverse ligament laxity [13,15].

Treatment of choice in Grisel syndrome is varied from conservative to surgical treatment. Until now, the management guideline on Grisel syndrome is still scarce. A review from Karkos et al. [15] stated that the more severe degree of subluxation and instability and Fielding type III and IV are associated with higher needs of surgical intervention. However, persistent neurological complication may ensue following surgical stabilization. Previous study reported persistent neurological complications in up to 15% patients underwent surgical intervention. Moreover, quadriplegia had been documented [15,16].

Orthosis is usually prescribed for children until they reach skeletal maturity. Less invasive techniques should be used on children to avoid postoperative deformity. Battiata et al. reported excellent results with manual reduction and Minerva cast in a 5-year-old [14]. Viscone et al. [12] reported similar results with nonsurgical treatment using manual reduction under general anesthesia and immobilization with Minerva cast in a 5-year-old girl. The study included patients with Fielding III, despite previous recommendations for surgical intervention, like our patient. The study concluded that surgery should be used as a last resort. Pilge et al. [17] reported reduced period of treatment with manual repositioning under general anesthesia and cervical spine immobilization in five patients. Moreover, no case of recurrence was observed within 6 months. That study is consistent with our finding that conservative treatment by manual repositioning and immobilization has more rapid recovery with painless and full range of motion.

However, surgical treatment such as release and fusion may be inevitable such as in persistent neurological symptoms. Another indication of surgical intervention include patient with Fielding type IV with subluxation and fractured dens axis or congenital dens-aplasia. Those patients are contraindicated for conservative treatment. Patient with delayed presentation of Grisel's syndrome also benefits from surgical intervention. Phillips et al. in their study found that around 60% pediatric patients with delayed treatment required fusion of C1-C2. Another study also added that the timing may be limited to three months [12,18].

4. Conclusion

Grisel's syndrome may appear to be worrisome deformity as a severe

wry neck with/without neurological deficit, nevertheless, it has excellent outcome following conservative treatment by closed reduction under general anesthesia and immobilization using Minerva cast for 6 weeks. Surgical treatment is indicated is very limited. Further study is needed to provide the best treatment option and absolute indication in both conservative and surgical treatment.

Declaration of competing interest

The authors declare that there is no conflict of interests regarding the publication of this paper.

Acknowledgements

Authors would like to give their biggest gratitude to the Department of Orthopaedic and Traumatology, Universitas Indonesia and Cipto Mangunkusumo Hospital for making this case report deliverable.

Funding

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

Ethical approval

Ethical approval was not required in the treatment of the patient in this report.

Informed consent

Written informed consent was obtained voluntarily from the parents for publication of this case and accompanying images.

Research registration

Case report does not need research registry.

Author contribution

SDALT was the attending surgeon and the guarantor, contributed in designing this study and final approval.

AH contributed in collecting and analysing data, and writing the manuscript.

RW contributed in collecting data and writing the manuscript.

Guarantor

SDALT is the guarantor.

Provenance and peer review

Not commissioned, externally peer-reviewed.

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