Case Report

Immediate postoperative resolution of syrinx post-C1/C2 fixation in an operated case of foramen magnum decompression for Chiari malformation: Is Goel's procedure a rescue surgery or a gold standard?

ABSTRACT

Chiari malformation (CM) is a common neurological disorder with foramen magnum decompression (FMD) as a commonly accepted treatment. The authors present a case of CM-1 wherein there was no radiological instability preoperatively and FMD was done as a treatment, after which the patient improved transiently only to deteriorate further. Atlantoaxial fixation was done as a second-stage procedure, after which the patient improved clinically and radiologically. The knowledge of this case and surgical entity should be borne in mind before the formulation of a treatment plan. It is important that the solution is to identify and treat the underlying pathology rather than to decompress and directly manipulate the tonsils.

Keywords: C1-C2 fixation, Chiari malformation, foramen magnum decompression, syringomyelia

INTRODUCTION

Chiari malformation (CM) is a common neurological disorder that is characterized by downward cerebellar tonsillar herniation through the foramen magnum. The prevalence of Chiari I malformation is one in 1000 births.[1] CM is often accompanied by basilar invagination (BI) and syringomyelia.[1-4] There are numerous hypotheses and causes which are proposed to explain the pathogenesis of CM.[5-9] The exact cause, however, is unclear or unknown. When there is no definite cause or when there is a bone anomaly associated with CMs, it is usually considered to be part of the whole complex and a birth defect. When syringomyelia is associated with it, which is not uncommon, it is often considered part of embryogenesis error. Anatomically, the syrinx formation is due to blockage of the cerebrospinal fluid (CSF) pathway due to herniation of cerebellar tonsils.[10] CMs are rarely associated with tumors in the back of the neck, hydrocephalus, or meningomyelocele. Very rarely, syrinx may be seen as a postoperative entity, especially after foramen magnum decompression (FMD) has been done. This suggests that there is a need to understand exact etiologies and CSF pathologies which need to be treated accordingly. It is important that the solution is to identify and treat the underlying pathology, rather than to decompress and directly manipulate the tonsils.[10-12] Authors present a case of CM-1 wherein there was no radiological instability preoperatively and FMD was done as a treatment after which the patient improved transiently only to deteriorate further.

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Submitted: 20-Dec-23 **Accepted:** 25-Dec-23
Published: 13-Mar-24

Access this article online	
Website: www.jcvjs.com	Quick Response Code
DOI: 10.4103/jcvjs.jcvjs_182_23	

How to cite this article: Chugh A, Punia P, Gotecha S, Gaud JA, Reddy R, Aziz RA. Immediate postoperative resolution of syrinx post-C1/C2 fixation in an operated case of foramen magnum decompression for Chiari malformation: Is Goel's procedure a rescue surgery or a gold standard? J Craniovert Jun Spine 2024;15:105-9.

CASE REPORT

A 38-year-old male patient was diagnosed with CM-1 [Figure 1] for which FMD with lax duroplasty [Figure 2] had been done as a treatment 6 months back. Currently, he presented with imbalance while walking for 20 days. After the first surgery, the patient transiently improved but deteriorated progressively to his current status (inability to walk independently). On examination, he has bilateral and LL weakness (UL > LL) with hypertonia (modified Ashworth grade 2) and brisk reflexes with severe gait disturbance (broad based).

The patient was thoroughly investigated and magnetic resonance imaging (MRI) craniovertebral junction (CVJ) suggested syrinx extending from the foramen magnum to C6, which was a new finding and was not previously seen before or immediately after the first surgery 6 months back [Figure 3].

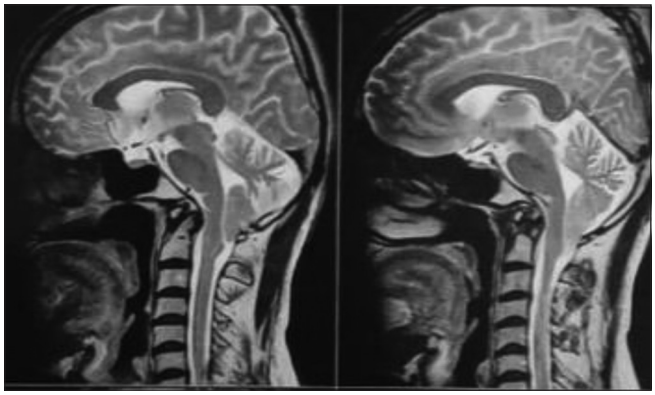

Figure 1: Preoperative magnetic resonance imaging craniovertebral junction with Chiari malformation-1 without syrinx

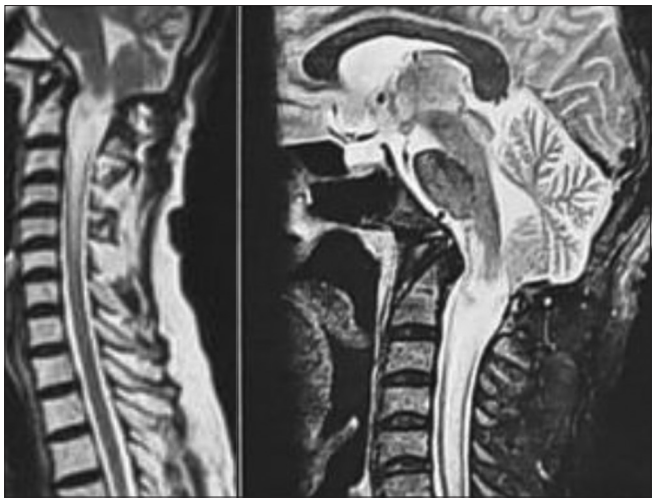

Figure 3: Postforamen magnum decompression magnetic resonance imaging cervical spine with craniovertebral junction suggesting Syrinx extending up to C6

Computed tomography (CT) CVJ suggested postoperative changes without obvious instability [Figure 4a-c].

The rest of the blood and other routine investigations were unremarkable.

The patient underwent atlantoaxial stabilization/fixation (C1 lateral mass and C2 pars screw) technique as first described by Goel and Laheri, Goel *et al*. [Figures 5-8].[1,3] In the immediate postoperative period itself, the patient reported considerable relief from tightness and improvement in handgrip. By the

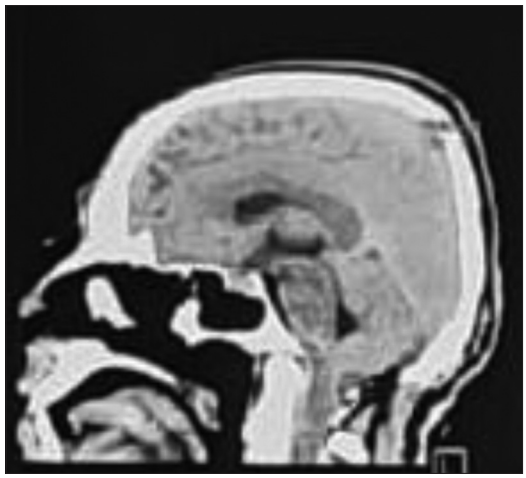

Figure 2: Postforamen magnum decompression computed tomography craniovertebral junction with postoperative changes

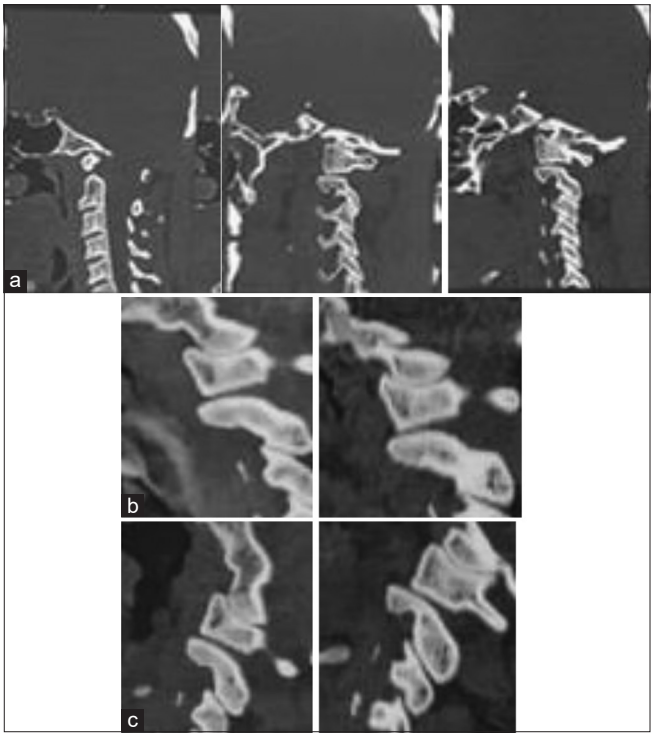

Figure 4: (a) Computed tomography (CT) Craniovertebral (CV) junction neutral, (b) CT CV junction extension, (c) CT CV junction flexion

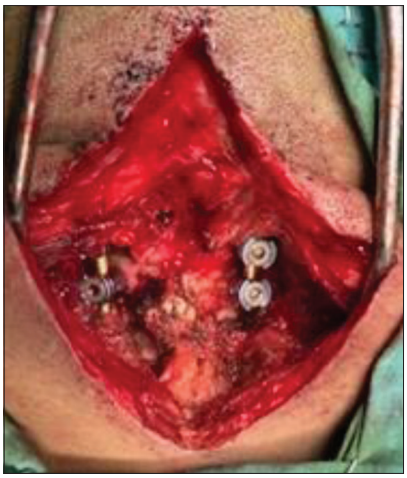

Figure 5: Intraoperative picture C1-C2 fixation

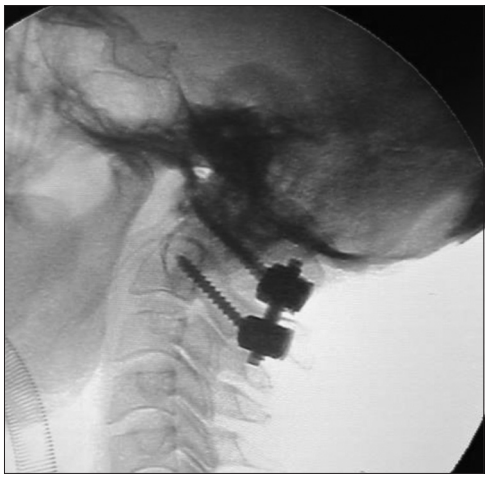

Figure 6: Intraoperative C1–C2 fixation lateral view

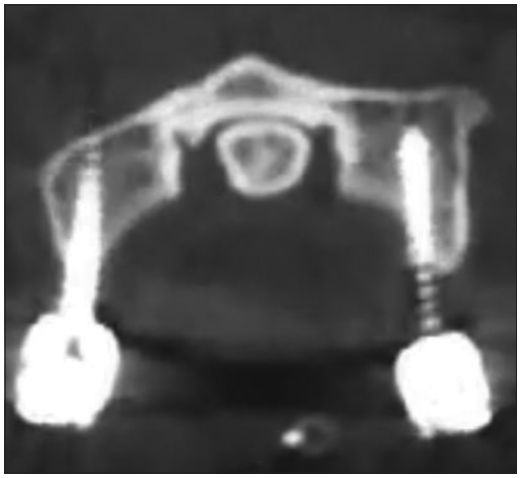

Figure 7: Postoperative CT C1 lateral mass fixation

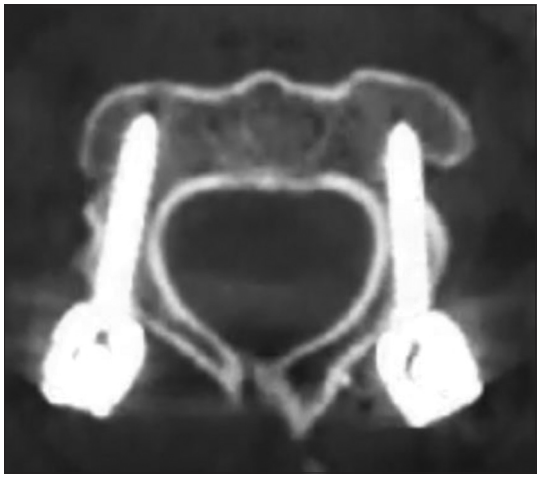

Figure 8: Postoperative CT C2 pars-pedicle fixation

7th postoperative day/before discharge, his handgrip and gait improved significantly and the patient was independently mobile. This rapid improvement prompted the surgical team to get an MRI CVJ done, which demonstrated beyond doubt the complete resolution of syrinx [Figure 9].

DISCUSSION

CM-1 is one of the commonly encountered entities in a neurosurgeon's practice. Few patients with CM may be asymptomatic and the spectrum of symptomatology varies from headache, imbalance, and motor weakness to giddiness, numbness, blurring of vision, and dysphagia. Our patient initially presented with dysesthesia, which was adequately relieved by FMD; however, he soon noticed tightness, which progressed rather fast, eventually limiting his ambulation. Each presenting symptom should be addressed and a detailed history along with examination and neuroimaging is important to understand and diagnose the pathology behind each symptom.[13-15]

MRI is considered the gold standard to diagnose CM and associated compressive or expansive pathologies, such as cerebellar tonsillar downward migration, hydrocephalus, syringomyelia, and spinal dysraphism. Dynamic CT craniovertebral (CV) junction is an important adjunct to MRI as it readily demonstrates detailed bony anomalies, including atlantoaxial dislocation, BI.[15] In our case, MRI promptly revealed syrinx, whereas CT CVJ was not suggestive of any obvious instability.

The primary aim of treatment of symptomatic CM with or without syrinx is to re-establish the CSF flow with neural decompression. The most common surgery performed to gain the aim is FMD with duroplasty. The success rate for the surgery is 70%–80%; however, there is often a subset of patients who need further management.[16-18] To recognize and address this subset, a better understanding of the etiopathogenesis of the disease needs to be developed, walking beyond the traditional concepts of stand-alone decompression surgeries. According to the newer concept forwarded by Goel in 2009, there are three types of atlantoaxial instability:[19-21]

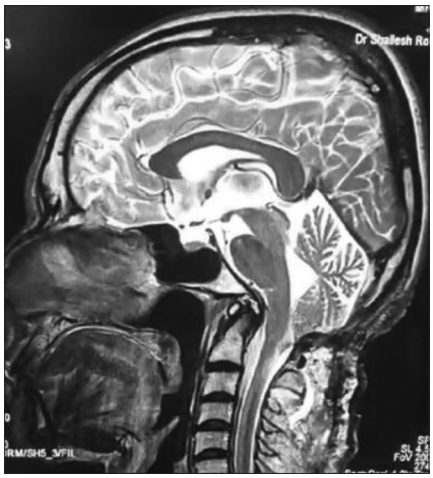

Figure 9: Postoperative C1–C2 fixation magnetic resonance imaging suggesting resolution syrinx

- Type 1: Anterior atlantoaxial instability
- Type 2: Posterior atlantoaxial instability
- Type 3: Central atlantoaxial instability (no radiologically demonstrable evidence of instability–can be worsened by traditional treatment of FMD).

Our patient belonged to type-3 instability wherein there was no change in atlantodental space and there was no neural or dural compression due to the odontoid process suggestive of obvious instability. Because of this, type 2 and 3 atlantoaxial facetal instability have been referred as "central" or "axial" atlantoaxial instability. In their study of 65 patients, Dr Goel perpetuated that CM, BI, and syringomyelia are not congenital malformations. Rather, all of these are clinical reflections of atlantoaxial joint instability and they may present as syringomyelia, BI, and CM, depending on the type and severity of joint instability. He further asserted that syringomyelia actually plays the role of a proverbial protective airbag for neural structures. The clinical progression of these patients usually follows a two-surgery path wherein after FMD, the neural structures that are under compression due to narrowed or obstructed CSF are relieved, resulting in transient clinical and radiologic improvement of the patient. However, this situation is temporary, as patients eventually return to their former clinical condition and may even worsen as a result of the underlying atlantoaxial instability. Hence, in this subset of patients, the primary aim should be to treat instability.[22,23]

It is clearly seen that until recently, no one gold standard surgical treatment has been effective for treating the whole CM complex; however, C1/C2 stabilization seems to do the trick. Our experience may be limited to this single case, but the fact that C1/C2 stabilization led to a reversal of syrinx points that it was not a matter of chance, especially when the concept has been backed by other clinical studies.

The recognition of predictive variables which help in determining which patients need FMD alone and which patient needs stabilization remains unclear. Whether C1–C2 fixation for all CM cases is really overkill, large numbers, and bigger studies are needed to ascertain the same. Till then, treatment with a proper explanation regarding C1–C2 fixation as a second stage procedure in patients undergoing FMD is required as being forewarned is being forearmed.

CONCLUSION

CM-1 is a simple pathology that is still poorly understood in terms of surgical treatment. Recognition of predictive patient factors to determine the right surgical procedure is the need of the hour. Authors stop just short of labeling C1–C2 fixation for all CM as a gold standard treatment because of a single case report and strongly suggest the need for a bigger patient study bracket to ascertain the same. However, the knowledge of this case and surgical entity should be borne in mind as being forewarned is being forearmed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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