Article published online: 2022-08-16

Case Report

Complex Chiari Malformation Presenting with Bulbar Symptoms in an Adult: Single-stage Posterior Fusion and Foramen Magnum Decompression: A Rare Case Report

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Abstract

Complex Chiari malformation (CCM) is a spectrum of congenital bony and soft tissue abnormalities, which includes Chiari 1.5 malformation, medullary kinking, retroflexed odontoid, abnormal clival-cervical angle (CXA), occipitalization of the atlas, basilar invagination, syringomyelia, and scoliosis. CCM usually manifests in the pediatric age group and is a challenging entity to treat. It requires detailed evaluation of craniometric indices to decide the appropriate surgical management. Patients with maximum perpendicular distance of dens to the line from the basion to the inferoposterior part of the C2 body (pBC2 line) of more than 9 mm and CXA <125° require a posterior fixation and will benefit from a single-stage posterior fusion and foramen magnum decompression (FMD). We report a rare case of CCM manifesting in a 32-year-old male with brainstem compression and bulbar symptoms. We could realign the craniovertebral junction with only a C1-C2 fixation by a modified distraction, compression, extension, and reduction technique and also relieve the neural compression by FMD and tonsillar resection in a single surgery with a good outcome. The authors find it to be an effective alternative to avoid the occipital fixation.

Keywords: *Adult, atlantoaxial joint fixation, Chiari malformation, decision-making, foramen magnum decompression, retroflexed odontoid, syringomyelia, vocal cord paralysis*

Introduction

Complex Chiari malformation (CCM) has been defined by Brockmeyer and Spader as a spectrum of bony and soft tissue abnormalities, which includes brainstem herniation through the foramen magnum (Chiari 1.5 malformation), medullary kink, retroflexed odontoid, abnormal clivocervical angle (CXA), occipitalization of the atlas, basilar invagination (BI), syringomyelia, and scoliosis.[1] These patients more often present with bulbar symptoms. CCM usually manifests in the pediatric age group and frequently requires posterior fixation in addition to the foramen magnum decompression (FMD). Manifestation of CCM in the adult age with brainstem compression and bulbar symptoms is a rare presentation. We report one such case where C1-C2 fixation by a modified distraction, compression, extension, and reduction (DCER) technique and FMD was done as a single-stage procedure.

Case Report

A 32-year-old male software engineer presented with unsteadiness when walking and hoarseness of voice for 1 year which worsened in severity over the past 2 months. Clinical examination revealed short neck, gait ataxia, left vocal cord palsy, left 12th nerve palsy, hypertonia of all four limbs, and severe posterior column impairment. Craniospinal magnetic resonance imaging and computed tomography (CT) of craniovertebral junction (CVJ) showed a CCM with BI, occipitalization of the atlas, rotational asymmetry of C1-C2 joint, retroflexed odontoid, maximum perpendicular distance of dens to the line from the basion to the inferoposterior part of the C2 body (pBC2 line) measuring 11.4 mm, CXA of 122.5°, 9 mm tonsillar descent, and cervicodorsal syrinx [Figures 1-3]. CT angiography revealed normal vertebral artery anatomy. ENT evaluation showed complete left side vocal cord palsy. In view of the progressive neurological

How to cite this article: Balasubramanian SC, Saphiya NN, Madan A, Mathews SS, Nair AR. Complex chiari malformation presenting with bulbar symptoms in an adult: Single-stage posterior fusion and foramen magnum decompression: A rare case report. Asian J Neurosurg 2020;15:132-5.

Submission: 26-11-2019 **Accepted:** 27-12-2019
Published: 25-02-2020

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Access this article online

Website: www.asianjns.org

DOI: 10.4103/ajns.AJNS_344_19

Quick Response Code:

worsening and associated BI with retroflexed dens and C1-C2 joint asymmetry, a single-stage posterior fixation and FMD was decided. The combined surgery was decided to avoid a secondary fusion procedure and also ensure that there is no further neurological deterioration which can occur if only a FMD was done. The patient was intubated with bronchoscopic guidance and positioned prone with three-point fixation. Midline skin incision made from external occipital protuberance to C2, muscle and subperiosteal dissection done and bilateral C2 dorsal ganglion was cut and C1-C2 joint space was exposed. C1 lateral mass and C2 pars-pedicle entry points were defined and screws were inserted under C-arm guidance (3.5 mm × 30 mm screw for C1 and 4.0 mm × 34 mm screw for C2). C1-C2 joint space was curetted and prepared. 5-mm cage on the right and 4-mm cage on the left were then inserted, and C1-C2 joint was distracted. Following this, compression of the C1-C2 joint was done and the entire construct was tightened. This was followed by FMD. Bony removal of dimension of 3 cm × 3 cm was done. After durotomy, dense arachnoid adhesions were observed and the left tonsil was seen to be significantly compressing the brainstem. Arachnoid adhesions were released and subpial tonsillar resection was done after which there was good cerebrospinal fluid outflow at the obex. Finally, lax duraplasty with fascia lata was done and glue was applied over the dural sutures. The patient was extubated on table and there was no intraoperative or postoperative complications. The patient had significant improvement in his gait and voice. Immediate postoperative CT showed good reduction of the BI and the pBC2 line was 8.6 mm which was 3 mm lesser than preoperative value of 11.4 mm [Figures 4 and 5a]. The scan repeated after 6 weeks showed good fusion at the C1-C2 joints [Figure 5b]. At 8-week follow–up, his mRS improved to 1 from preoperative score of 3.

Results

The novel surgical strategy employed by the authors has realigned the CVJ without the need for occipital fixation and effectively relieved the bulbar compression in the same surgery. The standard occipitocervical fusion surgery enables only *in situ* fixation of the atlantoaxial joints and it will not be effective to correct the rotational asymmetry at the atlantoaxial joint as seen with our case. Historically, in such cases with significant anterior compression by retroflexed dens, a transoral odontoidectomy used to be done. However, this procedure has high morbidity, and in recent times, anterior decompression procedure is done rarely and has given way for the posterior atlantoaxial fusion surgeries. In our case, we have used the distraction and compression technique for C1-C2 fixation by a modification of the DCER technique, and by combining it with FMD, we could effectively realign the CVJ, correct the joint asymmetry, and also achieve good neural decompression. An anterior decompression procedure and the need for a second fusion surgery has been avoided by doing this single-stage surgery.

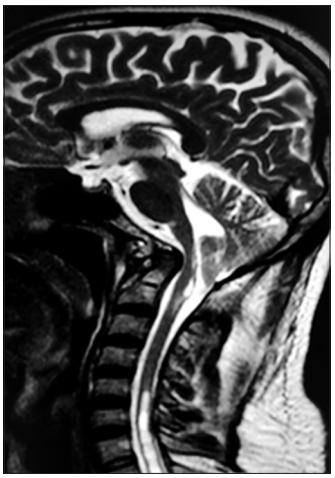

Figure 1: Preoperative magnetic resonance imaging showing the Chiari 1.5 malformation with syringomyelia

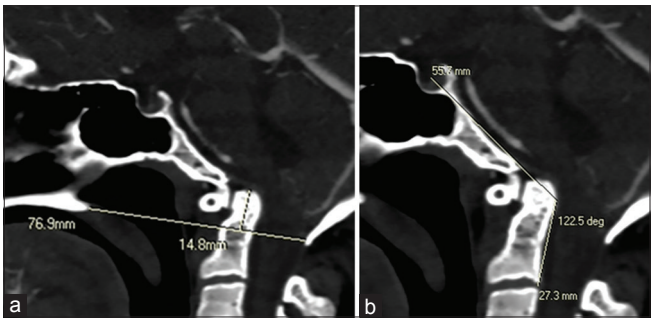

Figure 2: (a) Computed tomography craniovertebral junction showing basilar invagination. Tip of dens was 14.8 mm above the Chamberlain's line. (b) Computed tomography showing abnormal clivocervical angle 122.5°

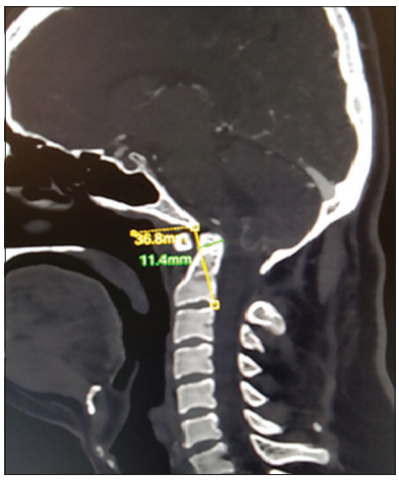

Figure 3: Computed tomography showing the preoperative pBC2 line measuring 11.4 mm

Discussion

CVJ anomalies are a broad spectrum of congenital bony and soft tissue pathology and include the bony segmentation defects, Chiari malformations (CMs), and BI. They may manifest alone or as a part of the syndromic conditions such as achondroplasia, Morquio, Marshall,

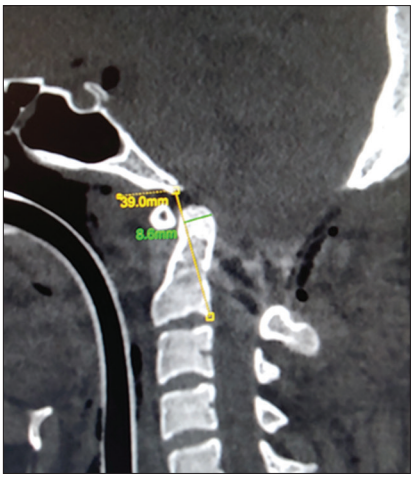

Figure 4: Postoperative computed tomography showing foramen magnum decompression and reduction in the pBC2 line (8.6 mm vs. preoperative 11.4 mm)

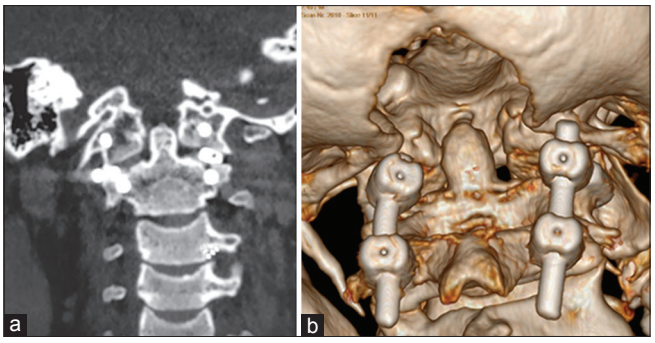

Figure 5: (a) Immediate postoperative computed tomography coronal view showing the spacers *in situ*. (b) Postoperative three-dimensional computed tomography at 6 weeks showing the fused construct with spacers and foramen magnum decompression

or Down syndrome.[2] There may be associated reducible or irreducible atlantoaxial dislocation (AAD). In 1896 Hans Chiari divided the CM into 4 types. The present classification of the CM has six types: Chiari 0, Chiari 1, Chiari 1.5, Chiari 2, Chiari 3, and Chiari 4. Chiari 1.5 specifically refers to patients with Chiari 1 malformation with additional elongation of the brainstem and fourth ventricle.[3]

Complex Chiari malformation: A new definition

Brockmeyer and Spader proposed a new category called the CCM which encompassed the spectrum of Chiari 1.5 malformation, retroflexed odontoid characterized by pBC2 distance >9 mm, CXA <125°, BI, assimilation of the atlas, and medullary kinking. It may be associated with hydrocephalus, syringomyelia, and scoliosis. They also concluded that patients with pBC2 of more than 9 mm and CXA <125° need both Chiari decompression and posterior fusion.[1]

Our patient fitted in the complex Chiari definition, the only difference being the adult age at presentation. Manifestation of CCM in adults with predominant bulbar symptoms is rare. We discuss in brief the history behind the management of CM and the factors which determine the choice of surgical approach.

Treatment options

The surgical treatment of CMs has seen a paradigm shift from only a bony FMD to combined fusion and decompression procedures. Anterior transoral decompression (microscopic or endoscopic odontoidectomy) is infrequently done these days and posterior fusion procedures with restoration of the joint alignment have gained the upper edge. These include occipitocervical fusion, C1-C2 fixation described by Atul Goel, and the DCER technique described by Sarat Chandra. The controversies as to which technique is superior continue till date, with each procedure having its own specific merits. More recently, Goel *et al*. proposed that the tonsillar herniation in CM is a protective mechanism due to the subtle or gross instability which occurs in all cases and hence C1-C2 fixations are required for all patients with CMs.[4] While the question of whether all patients require C1-C2 fixation remains debated, the spectrum of patients with complex bony abnormalities and rotational asymmetry always need some sort of posterior fusion procedure to restore the CVJ alignment.

It was Goel *et al*., who pioneered and popularized the C1-C2 fixation technique which leads to a new management strategy for CVJ abnormalities without the need for anterior decompression. They proposed the atlantoaxial joint distraction with direct lateral mass screw fixation and revolutionized the treatment of AAD with BI. This technique is now widely used, although there are issues with inadequate distraction and late recurrences which require revision surgeries or additional ventral decompression.[1]

The DCER which stands for distraction, compression, extension, and reduction is a novel technique pioneered by Sarat Chandra based on a 3-point lever principle and always includes occipital fixation along with C1-C2 fixation (joint remodeling with spacers or extra-articular distraction). The main indication is congenital cases of BI and AAD with occipitalized atlas. It is effective for even severe cases of BI with joint asymmetry.[2,5] However, restriction of the neck movement due to the occipitocervical fusion is the inherent disadvantage of this procedure.

Yerramneni *et al*. have described four cases of BI with irreducible AAD with the technique of C1-C2 fixation and combined it with the principles of DCER for reduction. They have got good results in all four cases and conclude that the combination of C1-C2 screw placement, followed by joint opening, spacer insertion, and subsequent distraction and compression at the screw heads has the potential to correct the alignment of the CVJ including the rotational component at the C1C2 Joints.[6]

Surgical decision-making in complex Chiari malformation

In our case, the complexity of the bony and soft tissue pathology fitting in the complex Chiari definition required a tailor made and targeted surgical approach to correct the specific abnormality. Management of a patient with CCM is challenging and requires a thorough preoperative assessment to evaluate the anatomical abnormalities and the biomechanical dynamics involved in the stability of the CVJ. Apart from the routine craniometric indices, we also have to quantify the retroflexion of odontoid and the clivus canal angle and analyze the C1-C2 joint orientation. Patients with retroflexed odontoid are more likely to have progression of ventral compression of the cord if only a FMD was done, and this subset of patients most often require a posterior fusion procedure. In our patient, the detailed preoperative evaluation of the craniometric indices revealed abnormality in both the orientation of the dens and the atlantoaxial joints, and a single-stage FMD and atlantoaxial joint fusion was the most appropriate choice.

The authors have used a similar technique as Yerramneni *et al*.[6] and have obtained a good outcome. The rotational asymmetry at the C1-C2 joint space was corrected using different sizes of spacers. We find this technique to be very effective when combined with FMD as a single-stage procedure and obviate the need for occipital fixation as well as an anterior decompression.

Conclusion

CCM in the adult is a challenging entity and requires detailed evaluation of the craniometric indices to decide upon the appropriate surgical technique. Patients with pBC2 of more than 9 mm and CXA <125° require a posterior fixation and will benefit from a single-stage posterior fusion and FMD. An adult patient with bulbar symptoms from a congenital CVJ anomaly with retroflexed odontoid is a rare presentation and we could realign the CVJ by a modified DCER technique and achieve neural decompression in a single-stage procedure with a successful outcome.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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