Case Report

Tethered cord and Chiari formation: Analysis of treatment in a relatively rare clinical situation

ABSTRACT

A 32-year-old male patient was operated for tethered cord associated with spinal lipoma. Further investigations showed the presence of basilar invagination, Chiari formation, and extensive syringomyelia. His neurological symptoms continued to worsen after the surgery, and over a 3-year period, he developed spastic quadriparesis and urinary retention and constipation. The patient was now treated by atlantoaxial fixation. Following the surgery, the patient improved in function in all four limbs and both the urinary and stool control. The presence of symptomatic Chiari formation in association with the tethered cord is a relatively rare clinical event. Surgical treatment of Chiari formation can result in a gratifying clinical recovery.

Keywords: Basilar invagination, Chiari formation, syringomyelia, tethered cord

INTRODUCTION

Chiari formation has been frequently identified in association with the tethered cord.[1] The general understanding has been that the tethering of the cord is the primary issue that results in "pulling" of the neural structures that include the cerebellum and the spinal cord caudally. In general, direct treatment for Chiari formation in such a twin pathological complex has only rarely been recommended or conducted. We report a rare clinical scenario wherein surgical treatment for Chiari formation resulted in relief from all presenting symptoms.

CASE REPORT

A 32-year-old male presented to another Institute with complaints of back pain and left lower limb weakness. Investigations performed at the time showed a low-lying tethered cord with a lipoma at the S1 region, Chiari formation, and syringomyelia [Figure 1a]. For these complaints, the patient was operated, and a detethering of the cord with partial excision of the lipoma and L5–S1 pedicle screw fixation was performed [Figure 1b]. The patient was well for a few months. After 10 months of surgery, the patient noticed that he had developed a foot drop and the weakness in his left lower limb had increased. This progressed to a feeling of stiffness and numbness in all his limbs. His symptoms gradually progressed over a period of 3 years, and he additionally developed difficulty in micturition and constipation. When he presented to us, he had spastic quadriparesis with difficulty in walking. The weakness was more pronounced in his left lower limb. He was able to walk with a stick with a limp. His grading was 3 when assessed by Goel's clinical grading scale.[2] He had decreased sensations to fine touch in his left leg and to pain and temperature in his right upper limb by 50%. He was catheterized and used laxatives to pass stools. Investigations focused on the

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craniovertebral junction showed basilar invagination with the assimilation of the atlas, Chiari formation, and cervicodorsal syringomyelia. Computed tomography scan showed that the atlantoaxial facets were in alignment (Goel Type C facetal instability) [Figure 1c-e].[3] Atlantoaxial fixation was now done as per the techniques described in 1994 and 2004 [Figure 1f and g].[4,5] Following the surgery, in the immediate postoperative phase, the patient improved significantly in all his complaints. The urinary catheter was removed 4 days after the surgery, and he regained normal urinary and bowel control. Furthermore, he observed improvement in his foot drop. At the follow-up of 10 months, apart from mild walking disability due to foot drop, there was no other neurological symptom. Magnetic resonance imaging repeated 3 months after surgery showed a reduction in the size of syrinx and evidences of fusion of the atlantoaxial joint [Figure 1h].

DISCUSSION

The presence of Chiari formation in association with the tethered cord is a well-known twin clinical entity.[1,6] The question is if the two otherwise discrete clinical entities have a pathogenetic relationship. The general understanding is that the tethered cord related to spina bifida, lipoma, thickened filum, and a range of other associated abnormalities is the primary issue that has a stretch effect on the spinal cord and the cerebellar tonsils. "Detethering" of the cord has been identified to be a definitive form of treatment for Chiari formation. Despite the fact that the progress of mechanical events looks obvious, some reports have suggested that both tethered cord and Chiari malformation could have independent origin meriting the need for two-stage surgical procedure for each clinical entity.[1] On the basis of their study, Roth identified that the tethered cord affects coordinated spinal column growth that can lead to both scoliosis and Chiari malformation.[7,8] Some authors have identified the presence of "tight" filum terminale as a causative factor for Chiari malformation and have demonstrated good surgical outcomes following sectioning of the filum terminale.[7] Clinical improvement has been identified in scoliosis, size of the syrinx, and neck pain following the surgery that is focused on tight filum terminale.[7,9-11]

In general, Chiari formation has been surgically treated by foramen magnum decompression. In the year 2013, we identified that Chiari formation is a secondary clinical event to primary atlantoaxial instability. On the basis of large clinical experience, it was hypothesized that atlantoaxial fixation is the treatment for Chiari 1 formation.[2] In the author's experience, none of the previously reported adult and pediatric patient cohorts with Chiari formation had associated tethered cord syndrome.

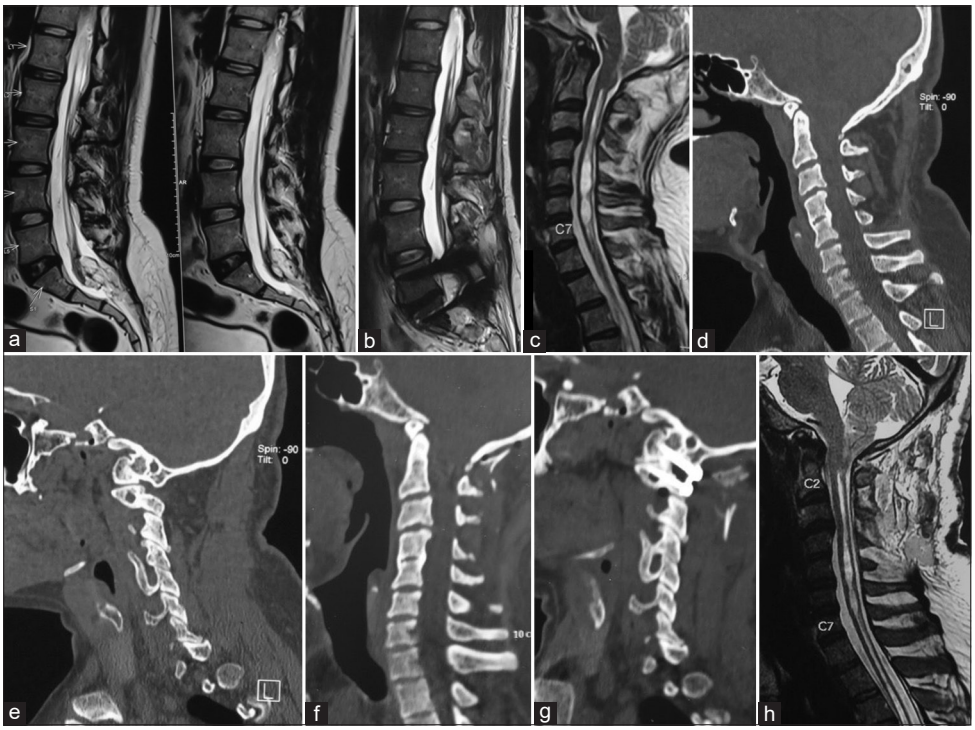

Figure 1: (a) T2-weighted magnetic resonance imaging showing tethered and low-lying spinal cord related to sacral lipoma. (b) Postoperative scan after detethering of cord and lumbosacral stabilization. (c) T2-weighted magnetic resonance imaging showing basilar invagination, Chiari malformation, and syringomyelia. (d) Computed tomography scan showing basilar invagination. (e) Computed tomography scan with the cut passing through the facets showing assimilation of the atlas. (f) Postoperative computed tomography scan. (g) Postoperative computed tomography scan with the cut passing through the facets showing the implant. (h) Postoperative T2-weighted magnetic resonance imaging (3 months after surgery) showing reduction of the size of the syrinx

The patient in the presented case had already been surgically treated for the tethered cord that was associated with a large intradural lipoma. In addition, segmental L5–S1 spinal fixation with screws and rods was done. Following an initial symptomatic improvement, the patient progressively worsened. Following the atlantoaxial fixation, the patient showed remarkable clinical recovery that started in the immediate postoperative period. The course of clinical events suggests the role of atlantoaxial instability in the case where tethered cord and Chiari formation are simultaneously identified. Considering that all the symptoms improved following atlantoaxial fixation surgery, one wonders if surgery for tethered cord could have been avoided. Furthermore, in the case scenario that is presented, the sequence of surgery could favor surgery for Chiari formation.

CONCLUSIONS

Atlantoaxial instability can be a major pathological entity in cases where Chiari formation and tethered cord are associated.

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Conflicts of interest

There are no conflicts of interest.

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