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Case Report

Acute traumatic presentation of Chiari I malformation with central cord syndrome and presyrinx in an infant

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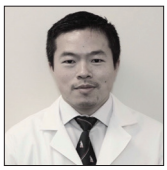

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ABSTRACT

Background: Chiari I malformation (CM-I) typically presents in late childhood and early adulthood. Often these lesions are asymptomatic and discovered incidentally. Patients typically present with tussive headaches and focal neurological findings, especially when associated with syringomyelia. Here, an 11-month-old child with a severely symptomatic CM-I required surgery (e.g., suboccipital craniectomy and C1/2 laminectomy) within the 1st year of life.

Case Description: An 11-month-old infant presented with acute bilateral upper extremity weakness following a ground-level fall. The magnetic resonance imaging of the cervical spine showed crowding at the craniocervical junction with 7 mm of cerebellar tonsillar herniation/descent, and swelling/edema of the cervical spinal cord with a presyrinx. The patient underwent an urgent suboccipital craniectomy and C1/2 laminectomy under intraoperative neuromonitoring; the motor evoked potentials in the upper and lower extremities partially recovered intraoperatively. One day postoperatively, bilateral upper extremity strength improved; 4 weeks later, he recovered full neurological function. The follow-up MR also showed complete resolution of the previously noted presyrinx.

Conclusion: Acute neurological deficits may occur in infants with CM-I who, following trauma, sustain the equivalent of a central cord syndrome. Neurosurgical evaluation with MR should prompt timely/appropriate surgical decompression (e.g., suboccipital craniectomy and C1/2 laminectomy).

Keywords: Central cord syndrome, Chiari I malformation, Posterior fossa decompression, Presyrinx, Syringomyelia

INTRODUCTION

Chiari I malformation (CM-I) is a congenital abnormality of the craniocervical junction (CCJ; frequency 0.1–0.5%). In an infant (i.e., <1 year of age) with CM-I, acute/severe neurological deterioration or even sudden death may follow even minor head/neck trauma resulting in a central cord syndrome.[7,9]

CASE REPORT

Clinical presentation

An 11-month-old boy presented with the acute onset of progressive bilateral upper extremity weakness. The previous evening, he had fallen backward, but had not sustained a loss of

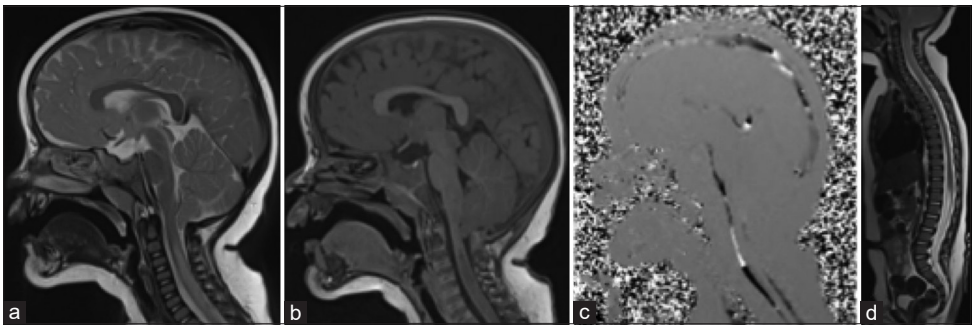

Figure 1: Chiari 1 malformation with presyrinx. Magnetic resonance imaging of the brain and spine demonstrate Chiari I malformation (CM-I) with tonsillar herniation 7 mm below the craniocervical junction. The cervical spinal cord is swollen from C2 to C6 with evidence of T2 (a) and T1 (b) signal elongation consistent with presyrinx. (c) Phase-contrast cerebrospinal fluid (CSF) flow study shows attenuation of CSF flow in the dorsal high cervical cord at the site of CM-I, which is preserved in the ventral spinal canal. (d) The remainder of the neuroaxis appeared normal without evidence of cavitation or syrinx.

consciousness or shown an focal neurological deficit. However, by the next morning, he showed diminished hand movement and was unable to grasp toys. By the time he was seen in the emergency department, he was unable to raise his arms above his shoulders. On examination, the cranial nerves were normal, but both upper extremities were flaccid, and he had absent upper extremity reflexes. Notably, he retained normal motion and reflexes in both lower extremities. Laboratory studies and infectious markers were negative, for example, white blood cell of seven and ESR of eight.

Radiologic studies

The magnetic resonance imaging (MRI) of the brain showed that the ventricles were of normal size without hydrocephalus [Figures 1a-d]. The brain and cervical MR studies, however, showed crowding of the craniocervical/cervicomedullary junction with cerebellar tonsillar descent of 7 mm below McRae's line [Figure 1a and b]. Phase-contrast cerebrospinal fluid flow studies revealed significantly reduced flow through the cerebral aqueduct [Figure 1c]. In addition, there was swelling of the cervical cord (on T1 and T2 images) from C2 to C6 consistent with cord edema (e.g., central cord injury) without bony or soft tissue trauma [Figures 1a and b].

Surgical management

Within 6 h of admission, after having received given 6 mg of dexamethasone, and using intraoperative neurophysiological monitoring (IONM: somatosensory evoked potentials and motor evoked potentials monitoring), the patient underwent a partial C1/2 laminectomy with suboccipital craniectomy extending to/through the foramen magnum. At surgery, there were thick fibrous bands of connective tissue that had to be dissected/excised until the superior aspect of C1 was visualized. Next, a C1 laminectomy was completed and decompression was taken laterally to the lateral masses; no duraplasty was performed. Intraoperatively, the motor evoked potentials partially recovered in both the upper and lower extremities.

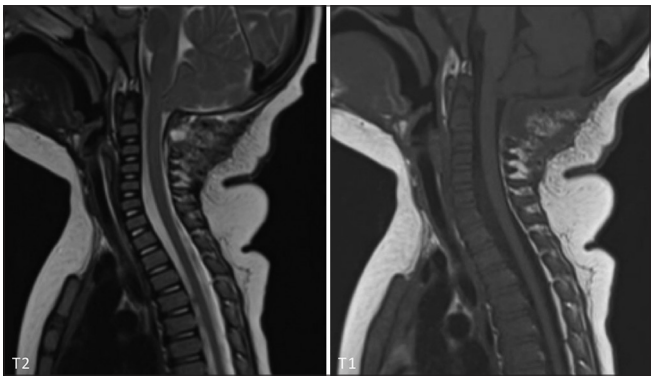

Figure 2: Resolution of presyrinx after surgery. One-month after bone-only posterior fossa decompression, magnetic resonance imaging of the cervical spine shows a similar degree of Chiari I malformation with tonsillar descent. However, the previously observed cervical spinal cord swelling and presyrinx have completely resolved. No other abnormality is observed.

Outcome

Postoperatively, the patient was extubated and was taken back to intensive care unit, moving all four extremities. He was treated with 3 mg of dexamethasone every 6 h for 3 days; this was tapered off over 5 days. On postoperative day 2, he was discharged home in a stable condition. Within 1 month, he had fully recovered. Further, the repeat MRI demonstrated total resolution of the cervical presyrinx, although the original CM-I remained stable without a change in tonsillar position [Figure 2]. Six months postoperatively, the patient remained neurologically intact.

Table 1: Reported cases of acute motor weakness following minor head/neck trauma in young children (<3 years of age) with Chiari I malformation.

Author	Age (Months) /Sex	Mechanism	Time to onset	Symptoms	Respiratory compromise	Imaging	Management	Recovery
Present	11 M	Fall backward	10 h	Paraparesis	No	Cord edema No syrinx	PF decompres/steroid	Complete
Massimi *et al.*[5]	12 M	Mild TBI	24 h	Hemiparesis	No	Cord edema No syrinx	PF decompres/steroid	Complete
Riviello *et al.*[6]	24 F	Fall from couch	2 h	Quadriparesis	Yes	No syrinx	Supportive	Partial
Bondurant and Oró[1]	27 M	Fall from porch	5 min	Quadriparesis	Yes	Cord edema No syrinx	Cervical collar	Partial
Vlcek and Ito[8]	30 M	Fall during handstand	Immediate	Paraparesis	No	No syrinx	Laminectomy	Complete
Massimi *et al.*[5]	30 M	Physical contact	12 h	Quadriparesis	Yes	Cord edema Syrinx	PF decompres+duraplasty	Partial
Erlich *et al.*[2]	33 F	Fall from bed	3 h	Paraparesis	Yes	Cord edema No syrinx	Supportive	Partial

M: Male, F: Female, PF decompress: Posterior fossa decompression, h: Hours, min: Minutes

DISCUSSION

The patient's presenting symptoms are consistent with traumatic central cord syndrome, with predominantly upper extremity weakness while sparing the lower extremities. Radiographically, the patient has CM-I with crowding of the CCJ, cervical cord edema, and a presyrinx.[3] Presyrinx is a clinical entity observed in patients with CM-I. Longitudinal follow-up without intervention suggests that the presyrinx progress to form syrinx, while surgical decompression of the Chiari often leads to resolution.[4] In this patient, it is unknown whether the presyrinx was a sequela to the neck injury, or whether the presyrinx was already present at the time of the traumatic event. Nevertheless, the rapid neurological recovery and resolution of the presyrinx following decompressive surgery are remarkable.

Occasionally, CM-I presents with acute and severe neurological deterioration or even sudden death following minor head/neck trauma.[5] In a retrospective review at a high volume pediatric neurosurgical center over a 19 year period, only six out of 498 CM-I patients (1.2%) had presented with acute neurological symptoms.[10] In another study, of 85 adult patients who presented with symptomatic CM-I, only three patients (3.5%) had the onset of symptoms that could be plausibly attributed to trauma.[9] The relationship between CM-I and trauma in young children is poorly understood. The precipitating head/neck trauma is often relatively minor and out of keeping with the degree of neurological deficit. Radiographic findings are variable and range from crowding of the CCJ with tonsillar descent to frank syrinx formation and intrinsic spinal cord signal abnormalities on MRI.

A review of the literature identified six other cases of CM-I in young children (<3 years of age) presenting with acute motor weakness following minor head/neck trauma [Table 1].[1,2,5,6,8] Their ages ranged from 11 to 33 months with a mean of 24 months, and two of the seven patients were female. In each case, the mechanism of trauma was minor and involved near-ground level incidents, which likely led to flexion/extension neck injury. None of the patients sustained a serious head injury or spine fracture. In all except 1 case, there was evidence of edema in the high cervical spine; however, only one patient was found to have a syrinx at the time of presentation. Interestingly, three of the seven patients made a full neurological recovery and all three of these patients had undergone surgical decompression. Nevertheless, due to the infrequent cases, the correct management is unknown and neither is the appropriate timing of surgical intervention. However, in young patients who present with acute neurological deficits, surgical decompression of the CM-I appears to be well tolerated and improve clinical symptoms with stabilization or resolution of associated cord edema/presyrinx.

CONCLUSION

CM-I usually presents with indolent symptoms and subtle neurological signs, and seldom requires surgical management in infants. However, it can occasionally present following minor trauma with acute neurological deficits. In such cases, urgent referral for neurosurgical evaluation is warranted and early decompressive surgery should be considered.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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