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Case Report

Intractable hiccup caused by syrinx in Chiari type I malformation. Two cases report

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ABSTRACT

Background: Intractable hiccups (IH) due to syringomyelia or syringomyelia/syringobulbia associated with Chiari type I malformations (CMI) are extremely rare. Here, we present two patients who presented with IH; one had a CMI with syringomyelia/syringobulbia, and the other, with CMI and syringomyelia.

Case Description: The first patient was an 18-year-old female who presented with IH attributed to a holocord syrinx and syringobulbia involving the right dorsolateral medulla. The second patient was a 22-year-old female with a C3-5 syringomyelia. Both patients successfully underwent foramen magnum decompressions that improved their symptoms, while subsequent magnetic resonance studies confirmed shrinkage of their syringobulbia/syringomyelia cavities.

Conclusion: IH was due to cervical syringomyelia/syringobulbia in one patient and cervical syringomyelia in the other; both were successfully managed with foramen magnum decompressions.

Keywords: Chiari type I malformation, Intractable hiccups, Lateral medullary syndrome, Syringobulbia, Syringomyelia

INTRODUCTION

Intractable hiccups (IH) are rarely due to Chiari I malformation (CMI) in conjunction with cervical syringomyelia/dorsal medullary syringobulbia and/or cervical syringomyelia.[1,2] Here, we present two patients with IH, respectively, attributed to CMI with cervical syringomyelia/syringobulbia, whereas the other had cervical syringomyelia alone; both were successfully treated with foramen magnum decompressions.

CASE REPORT

Case 1

An 18-year-old female presented with 6 months of hiccups, and difficulty writing due to the distal right upper extremity muscle atrophy. The magnetic resonance imaging (MRI) showed

a CMI with a holocord syrinx and a slit-like cavum on the dorsal lateral side of the right medulla (syringobulbia) [Figure 1a]. Two weeks following foramen magnum decompression and C1 laminectomy without duraplasty, her cough, headache, and IH rapidly improved. One year postoperatively, the magnetic resonance (MR) showed that the spinal cord syringomyelia and syringobulbia (medullary oblongata) had markedly regressed [Figure 1b]. Two years postoperatively, she remained symptom-free, and the syringobulbia cavity on the MRI had been further reduced.

Case 2

A 22-year-old female presented with 2 months of hiccups, and 3 months of dizziness accompanied by the right upper extremity numbness. The MRI showed a CMI and a cervical syrinx from C3 to 5 [Figure 1c]. One month following foramen magnum decompression and a C1 laminectomy without duraplasty, her symptoms rapidly improved. At 10 postoperative months, she was asymptomatic, and the follow-up MR documented no regrowth of the syrinx.

DISCUSSION

Symptoms and pathophysiology of CMIs

CMI is the most common etiology of cerebellar tonsil herniation at the craniovertebral junction. Accompanying syringomyelia is diagnosed in 20–50% of these patients. Clinical symptoms of CMI with syringomyelia include cough, headache/occipitalgia, gait disturbances/ataxia, dissociated sensory disorders, dysphagia, and paresis of the extremities.[2]

Pathophysiology of hiccups

Hiccups are defined as intractable when they; persist for more than 24 h, when the rate increases from 40 to 100/min, and/or when they become refractory to usual methods of treatment.[1,4,7] IH is typically attributed to involuntary spasmodic contractions of the diaphragm accompanied by the sudden closure of the glottis; this produces a classic click sound (i.e., singultus or hiccup). Here, we were only able to identify five articles citing six patients who presented with IH [Table 1].[1,2,5,6,8]

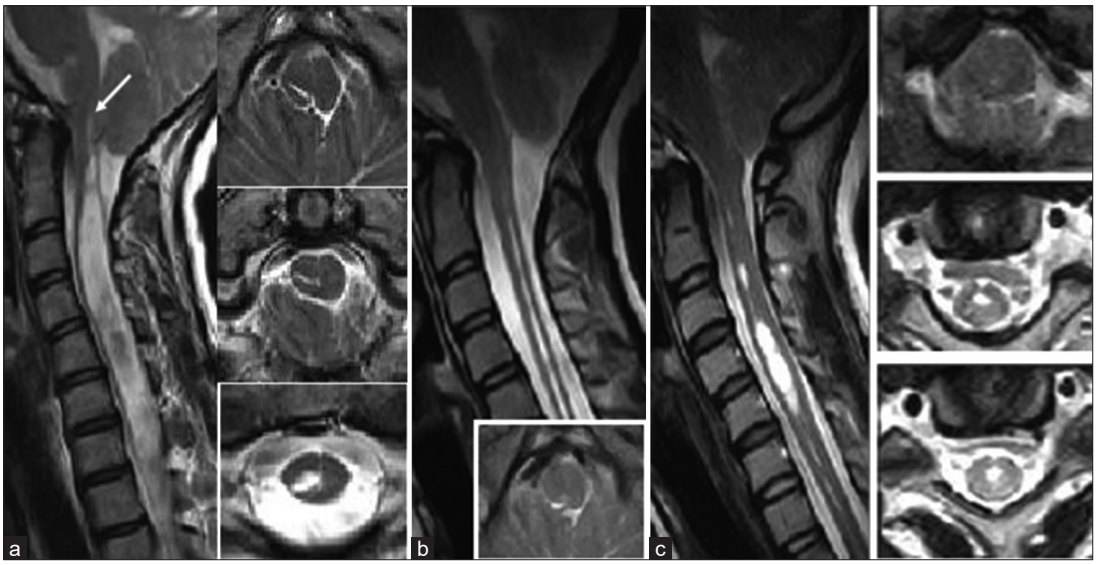

Figure 1: (a) Case 1: Preoperative magnetic resonance imaging (MRI) scans of the cervical spine. T2-weighted MRI scan in the sagittal plane, showing a Chiari type I malformation (CMI) and a holocord syrinx with syringobulbia (arrow). An axial MRI scan was obtained at the foramen magnum level, demonstrating the slit-like syrinx in the right dorsal region of the lower medulla. (b) Case 1: Postoperative MRI scans of the cervical spine obtained 3 months after surgery. T2-weighted MRI scan in the sagittal plane, demonstrating decompression of medulla oblongata and cerebellar tonsils at the foramen magnum. The syringobulbia has collapsed. The cervical syringomyelia is slightly decreased in size but remains and (c) Case 2: MRI scans of the cervical spine. T2-weighted MRI scan in the sagittal plane, showing a CMI and a syrinx from the high cervical to the C3-5 level. Axial MRI scan was obtained at the foramen magnum and C3-5 level, demonstrating the syrinx in the mid-ventral region.

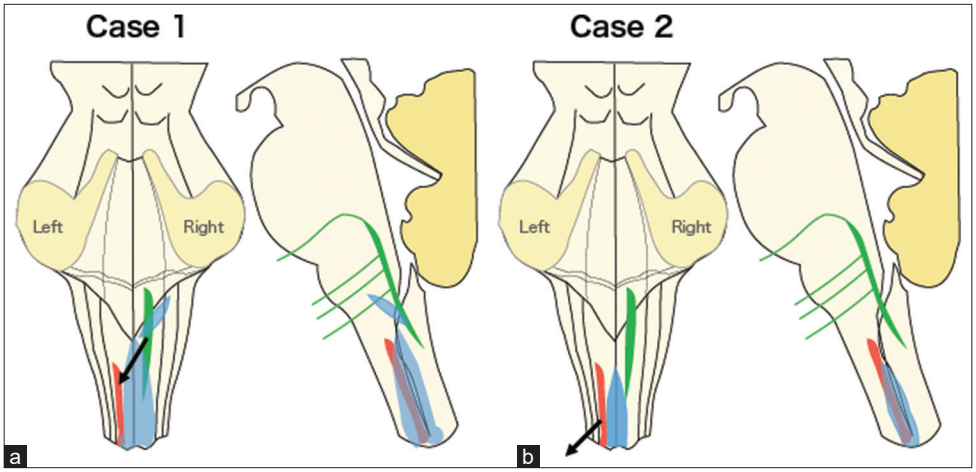

Figure 2: The schema of pathophysiological mechanisms for hiccups due to syrinx with Chiari type I malformation (CMI). (a) Case 1: CMI with syringomyelia and syringobulbia (blue) and (b) Case 2: CMI with syringomyelia (blue). "Hiccup center" accessory nucleus (red) in the upper spinal cord (C3-C5), nucleus solitarius (green) in the medulla oblongata near the respiratory center. The stimulation to hiccup reflex (black arrow).

Table 1: Summary of cases of Chiari I malformations with hiccups.

Author	Age/Sex	Syringomyelia	Syringobulbia
Seki *et al*.[5]	27/M	C2-7	Right dorsal
Shankar *et al*.[6]	22/M	C2-T7	Right dorsal
Present case 1	18/F	Holocord	Right dorsal
Amirjamshidi *et al*.[1]	21/M	Holocord	No
	19/M	C3-	No
Vanamoorthy *et al*.[8]	22/M	C1-4	No
Chen *et al*.[2]	34/M	C2-4	No
Present case 2	22/F	C3-5	No

M: Male, F: Female, C: Cervical, T: Thoracic

CM1 with hiccups

We have added two patients to the six cases previously reported who presented with both IH and CMI [Table 1].[1,2,5,6,8] The six patients from the literature combined with the two from this study averaged 23.1 years of age, all exhibited cervical syringomyelia, whereas an additional three of eight patients also had syringobulbia (37.5%).[5,6] Moon *et al*. additionally reported patients as presenting with Wallenberg syndrome along with hiccups[3] [Figure 2].

CONCLUSION

Two young females, 18 and 22 years of age respectively, presented with IH attributed to CMI/syringomyelia and CMI/syringomyelia/syringobulbia; both were successfully treated with foramen magnum decompressions.

Declaration of patient consent

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

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Nil.

Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The author(s) confirms that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

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