Yawning as a presenting symptom of Chiari malformation Type I: report of 2 cases

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Yawning is thought to be a behavior regulated by the brainstem. Although excessive yawning has been reported in brainstem strokes, demyelination, and tumors, the cases presented here are the first reports of excessive yawning in patients with Chiari malformation Type I (CM-I). The authors believe that brainstem compression at the craniocervical junction and ensuing edema were implicated in this curious symptomatology. They describe excessive yawning as a presenting feature of CM-I in 2 adolescent females. The presentation was acute in the first case and more chronic in the second. Both patients underwent foramen magnum decompression, which resulted in complete cessation of the excessive yawning.

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KEY WORDS Chiari; yawning; brainstem; foramen magnum; decompression

Case Reports

Case 1
History and Examination
A 12-year-old girl whose twin sister had undergone foramen magnum decompression for CM-I was referred to the pediatric neurology department with a 2-week history of gait disturbance and right-sided weakness. While undergoing outpatient MRI, she was noted to be lethargic with persistent yawning. She was referred to the on-call neurosurgical service for urgent review.

Magnetic resonance imaging demonstrated tonsillar descent, an extensive syringomyelic cavity in the cervicothoracic cord, and signal change in the ventral and central medulla without syringobulbia (Fig. 1).

Operation
Four days later, she underwent foramen magnum decompression (bony decompression with arachnoid-sparing opening of the dura mater).

Postoperative Course
She made a good recovery with residual but improving mild right upper limb weakness. Her yawning and lethar-
gy ceased postoperatively, and at the 36-month follow-up she remained very well.

Case 2

History and Examination

A 15-year-old girl presented to clinic with a 2-year history of increasingly frequent headache following a minor trauma. In addition, she described an 8-month history of difficulty swallowing and pooling of saliva in the mouth and throat. A barium swallow had been performed and failed to provide a diagnosis. She also reported a 4-month history of “muffled speech” and difficulty articulating her words.

Throughout the clinic appointment, frequent yawning was noted. On further questioning, the patient and her mother agreed that yawning had been occurring with increasing frequency, even preceding the symptoms described above. They could not remember the onset of the frequent yawning. On average, the yawning episodes would occur once every 10 minutes and would last for just 1 or 2 minutes with persistent yawning each time. She denied feeling lethargic, and there was no clear diurnal variation, although she had not been observed as an inpatient during sleep. Yawning was more frequent when she was suffering from headache. On examination, she was neurologically intact.

Magnetic resonance imaging of the neuraxis revealed a CM-I with retroversion of the odontoid process causing severe crowding at the foramen magnum and an associated cervicothoracic syrinx with faint edema in the medulla (Fig. 2).

Operation

She underwent expedited foramen magnum decompression (bony decompression with dural opening and sparing of the arachnoid).

Postoperative Course

Postoperative recovery was uncomplicated, and she was discharged home within a week. She reported significant improvement in her headache, improved speech due to reduced pooling of saliva, improved swallowing, and cessation of the excessive yawning. At the 30-month follow-up she remained well with no further symptoms.

Discussion

Hans Chiari (1851–1916), an Austrian professor of anatomy and pathology in Prague, is credited with the first descriptions (1891 and 1896) of 4 types of hindbrain malformations (CM-I, -II, -III, and -IV). The exact definition of each has evolved since.

Two other entities have also been described. Chiari malformation Type 0 is characterized by syringomyelia in the absence of hindbrain malformation and responds to surgical intervention in much the same way as CM-I. Chiari malformation Type 1.5, on the other hand, is characterized by descent of the tonsils, as well as the brainstem and fourth ventricle, through the foramen magnum but in the absence of a myelomeningocele.

Although it used to be widely accepted that CM-I presents in the 2nd or 3rd decade of life, the disorder is being diagnosed more frequently in the younger population since the advent of MRI. The most common presenting symptom is occipital and/or cervical pain that is often caused or exacerbated by straining; however, the pain may have a more constant dull element. Other presenting symptoms are, in order of their frequency, limb weakness, numbness, dissociated sensory loss, unsteadiness, and cranial nerve palsies. Other much less common symptoms such as hiccoughs and fainting have also been reported. To our knowledge, these are the first reported cases of CM-I presenting with yawning.
The function of yawning remains a topic of debate. Some believe that it increases vigilance in states of low arousal; others, that it serves a role in social communication and in synchronizing the physiological and behavioral state of a group. The “deep inhalation” phase of yawning may serve as a respiratory reflex designed to increase oxygenation in states of hypercarbia, although experimental evidence does not support this theory. It is, however, accepted that yawning is a common human response to states of fatigue or boredom. It consists of 3 phases: The first, or “inspiratory phase,” involves wide mouth opening and deep inspiration. The nostrils and soft palate are elevated, the nasopharynx dilates, the tongue moves posteriorly, and the vocal cords abduct. At the peak of inspiration there are associated facial movements and often arm and/or trunk extension. The second phase, or “acme,” is where inspiration is maintained. Lacrimation and salivation may occur in tandem with eye closure. Eustachian tube closure precipitates transient hypoacusis. Maximal arm and/or trunk extension can alter proprioception. The final phase, or “expiratory phase,” involves rapid exhalation followed by a brief apneic episode.

Yawning occurs in neonates with anencephaly, indicating that the brainstem plays a key role in this automatism. Cattaneo et al. concluded in their review of the published literature that the circuit responsible for yawning involves the hypothalamus, midbrain, and reticular formation of the pons and medulla. Although dopamine and oxytocin play a central part, it seems that other substances are also involved, including nitric oxide, acetylcholine, orexins, and serotonin as well as opioids. Effector centers that generate the yawn include the bulbar respiratory center; the motor nuclei of cranial nerves V, VII, X, and XII; the phrenic nerve; and the nerves supplying the accessory muscles of respiration.

Pathological or excessive yawning is defined as a “compulsive, repetitive action that is not triggered by appropriate stimuli such as boredom or fatigue.” A literature search for “pathological yawning” showed it has been observed in various neurological pathologies including basal ganglia disorders, multiple sclerosis, progressive supranuclear palsy, infarcts, and tumors but not in the context of CM-I.

Conclusions

The anatomical pathways controlling yawning are not fully understood, but there is clear evidence that the brainstem plays a key role. Excessive yawning has been reported in brainstem stroke, tumor, and demyelination, but these are the first reports of excessive yawning in patients with CM-I. The presence of brainstem edema on imaging is unusual in patients with CM-I. We believe that the weakness of the limbs, lower cranial nerve weakness, and pathological yawning in our patients resulted from compression of the brainstem at the craniocervical junction with ensuing edema.

Although disturbed sleep was not reported in either of the featured cases, the well-known relationship between CM-I and sleep disorders (particularly sleep apnea syndrome [SAS]) may have contributed to the yawning. A literature search was performed to see if a relationship between SAS and excessive yawning was documented, but no reports were found to support this. However, neither of the patients was observed during sleep as we performed expedited foramen magnum decompressions on both, which precluded us from being able to perform sleep studies.

These cases highlight the probable involvement of the brainstem in yawning and the potential for CM-I to present with excessive yawning. We propose that when this occurs, urgent decompression is indicated to relieve brainstem compression.

References

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Author Contributions

Conception and design: Zebian. Acquisition of data: Zebian, Hogg, Fu. Analysis and interpretation of data: Zebian, Hogg, Fu. Drafting the article: Zebian, Hogg, Fu. Critically revising the article: Zebian, Sivakumaran, Stapleton. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Zebian.

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