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What is This?
Chiari I Malformation: Patient Report and a Mini Review

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Introduction

Chiari I malformation refers to the downward herniation of the cerebellar tonsils through the foramen magnum. It may occur in pediatric patients as an incidental finding or may present with varied and unusual clinical symptoms. The case of a 17-year-old female who presented with a 2-year history of chronic back, left hip, and left leg pain is discussed. She was subsequently diagnosed with borderline Chiari I, pseudotumor cerebri, and HAIR-AN syndrome.

Case Presentation

A 17-year-old white female presented with a 2-year history of back, left hip, and left leg pain. The pain initially developed in the low back. It gradually worsened and extended to her left hip and leg. She rated her daily pain as 7 to 8 out of 10 with occasional exacerbations to 10 out of 10. Eight months later she developed new onset headaches. She had 3 to 4 headaches daily with occasional photophobia and intermittent diplopia. She subsequently developed a 1-year history of ataxia. The pain eventually prevented her from attending school or participating in her usual physical activities.

Prior evaluations by orthopedics, neurology, immunology, and rheumatology yielded a differential diagnosis that included lupus, ankylosing spondylitis, and left sciatic neuritis. She tried several medications including non-steroidal anti-inflammatory drugs (NSAIDs), Plaquenil, Relafen, Elavil, Zonaflex, prednisone, Celebrex, Mobic, Vioxx, and ketoprofen with limited relief. Her past medical history was negative. She denied use of tobacco, drugs, alcohol, sexual activity, and physical or sexual abuse.

Her previous workups comprised: negative magnetic resonance imaging (MRI) of lumbar spine; whole body bone scan consistent with inflammatory disease involving bilateral sacroiliac joints, knee joints, tarsals of both feet and ankle joints, both shoulders and hip joints, left hip and shoulder greater than the right; negative head CT; negative electromyography and nerve conduction study and a cervical-spine MRI demonstrating cerebellar tonsillar ectopia; a normal complete blood count (CBC) and differential, elevated erythrocyte sedimentation rate (ESR) (40–60), negative rheumatoid factor; positive anti-nuclear antibody (ANA) (1:160 with a homogeneous pattern); normal thyroid studies; negative antibody for H. pylori; and negative hepatic panel.

Upon presentation to our clinic, her physical examination was significant for obesity, depressed mood, an abnormal gait characterized by limping and bearing weight on toes of left lower extremity, facial acne, acanthosis nigricans involving her neck, axillae, and antecubital fossae, blurred right optic disc, and mild nonpitting edema over left ankle. She complained of left knee, hip, and leg pain with light touch and resisted internal and...
Chiari I Malformation

Chiari I malformation is a congenital downward herniation of the cerebellar tonsils through the foramen magnum. This condition is characterized by symptoms and signs that are related to compression of the brainstem and cerebellar structures. The clinical manifestations of Chiari I malformation can include headache, fatigue, dizziness, and motor and sensory deficits. The diagnosis is typically made using magnetic resonance imaging (MRI) of the cervical spine. Treatment options include observation, pain management, and surgical intervention in cases of symptomatic compression or progressive symptoms.
The symptoms of Chiari I malformation can often be vague and ambiguous in children, leading to misdiagnosis. A review of the records of 11 children who underwent suboccipital decompression for symptomatic Chiari type I malformation revealed that presenting complaints included neck pain, scoliosis, back pain, torticollis, motor dysfunction, and apnea.

Stevens et al suggested that the clinical symptoms ascribed to cerebellar ectopia could be related to the severity of the malformation and that the operative outcome could be related to the morphological findings. They determined that patients with the most severe cerebellar malformation, defined as descent of the cerebellar tonsils to or below the axis, had disabling ataxia and nystagmus more frequently. Limb weakness and muscle wasting were more frequently seen in patients with brainstem compression. Postoperative outcome was less favorable in patients with severe cerebellar ectopia (12% improved, 69% deteriorated) than in those with minor ectopia (50% improved, 17% deteriorated). Patients with distended cervical syrinx had a more favorable outcome than those without.

Loder et al studied the association of scoliosis with Chiari I malformation. They retrospectively reviewed 30 children with scoliosis secondary to Chiari I from three centers. Syringomyelia was present in 87% of the patients. The scoliosis was thoracic in 25, thoracolumbar in 3, and lumbar in 2; 18 curves were right and 12 were left. A positive correlation was noted between cervical lordosis and thoracic kyphosis. When cervical lordosis is greater than zero degrees or thoracic kyphosis is greater than 40 degrees (Cobb method), the clinician should suspect the presence of Chiari I malformation with or without syringomyelia. Compared to adolescent idiopathic scoliosis, the patients with Chiari I and associated scoliosis had more left curves (40% vs. 0%), were more like boys (37% vs. 8%), and were younger (11.3 years vs. 14.2 years). Steinbok also demonstrated that scoliosis is an important and common finding in children with syringomyelia associated with a Chiari I malformation. Progressive scoliosis was found in approximately 30% of such children.

Patients with Chiari malformations may also present with vestibular system symptoms, including ataxia, nystagmus, or vertigo. Patients with advanced symptoms may demonstrate oculomotor dysfunction, central vestibular nystagmus, abnormal vestibular visual interaction, and abnormal tilt suppression of postrotatory nystagmus.

Pseudotumor cerebri associated with Chiari I malformation has been reported in two 12-year-old patients. Although papilledema has been reported occasionally, a causal relationship has not been established and the mechanism for the increased pressure is not well understood. The clinical distinction between Chiari I malformation and pseudotumor cerebri is sometimes difficult, as patients with pseudo tumor cerebri and Chiari I malformation usually become symptomatic in young adulthood and are usually women. However, patients with pseudotumor cerebri are usually obese, whereas patients with Chiari I malformation are not necessarily obese.

Treatment and Management

Considerable debate exists about when surgery is indicated and which surgical options are best for management of Chiari I malformation with and without syringomyelia. There are at least 15 different methods and about 40 different operative methods. The surgical treatment for Chiari I malformation can stabilize or slightly improve the symptoms of syringomyelia and relieve the symptoms of brainstem compression.

Type A patients had predominantly central cord symptoms and type B patients had primarily brainstem or cerebellar compression symptoms. The principal surgical procedure consisted of decompression of the foramen magnum, opening of the fourth ventricular outlet, and plugging of the obex. Significant improvement in preoperative symptoms and signs was observed in 45% of type A patients compared to 87% of type B patients. The presence of syringomyelia implied a less favorable outcome.

The main benefit of surgical management in patients with Chiari I malformation without or with syringomyelia was prevention of disease progression. Overall, surgery improved both symptoms and signs, but the improvement in symptoms was more marked. There was a positive correlation between improvement in hydrosyringomyelia and the improvement in signs and symptoms.
Outcome and Prognosis

The mean interval between onset of symptoms and operation was shorter in the children (3 years 6 months vs. 7 years 1 month); preoperatively adults had more severe symptoms; and postoperatively seven of eight pediatric patients improved and one stabilized whereas two of five adults improved, one stabilized, and the disease progressed in two. Additionally, the preoperative abnormal cerebrospinal fluid flow at the craniovertebral junction and to-and-fro movement in the syrinx improved.16

Current opinion on the treatment of syringomyelia and Chiari malformation supports a standard bony and dural decompression of the foramen magnum region with modifications designed to maximize the restoration of cerebrospinal fluid circulation across the foramen magnum is a safe, effective operative treatment for Chiari I malformation in children.17-20

Summary

Children with Chiari I malformation present with varied and unusual presentations that may lead to missed or misdiagnosis and delayed treatment. Pediatric patients with chronic pain may benefit from evaluation for Chiari I malformation to ensure early treatment and prevention of progression of the disease. Chiari I malformation in patients with chronic pain may not be an incidental finding.

Acknowledgments

Written consent was obtained from the patient or their relative for publication of study.

REFERENCES


