## Title

Chiari I Malformation with Syringomyelia as a Cause of Nausea and Vomiting



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**CLINICAL VIGNETTE**

# Chiari I Malformation with Syringomyelia as a Cause of Nausea and Vomiting

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***Case Report***

A 25-year-old male with nausea and vomiting presented for outpatient evaluation. He complained of awakening every morning with nausea and then repeated vomiting. He reported 15-20 episodes of small volume vomiting each day. Initially his symptoms were worse in the mornings, but then evolved to occupy the entire day. The symptoms started five years prior to this evaluation. His weight had fluctuated within a five-pound range. He had previously been seen and undergone extensive evaluation at outside facilities. These tests included EGD with random biopsies, colonoscopy with random biopsies, CT scan of the abdomen and pelvis, small bowel follow through and gastric emptying study. All of these studies were unremarkable and his symptoms were not relieved with a trial of omeprazole, odansetron, or metoclopramide. The patient had no other significant past medical history and family history and social history were unremarkable.

On physical exam he had a temperature of 98.9 F, pulse of 91, blood pressure of 119/80 and weight of 175 pounds. There were no significant positive findings on physical exam. There were no laboratory studies available for review at the time of initial consultation. Further diagnostic testing was ordered, including barium esophagram, celiac antibody panel, and MRI of the brain to rule out central causes of his symptoms. The barium esophagram was normal and celiac serologies were negative. The brain MRI showed a Chiari malformation with the cerebellar tonsil tips extending approximately 2 cm below the level the foramen magnum; a small syrinx was also visible in the upper cervical cord. This study was followed by MRI of the cervical spine which showed a Chiari type I malformation with cervicomedullary junction anomaly, the medulla was deformed at the foramen magnum, and a syrinx was seen from C2 to C6.

Based on these findings the patient was referred to Neurosurgery for further discussion of treatment options for the Chiari I malformation with syringomyelia. He ultimately underwent surgery, which included[1]: suboccipital craniectomy and C1 laminectomy with intradural exploration, shrinkage of cerebellar tonsils, decompression of the spinal cord and brainstem and pericranial duraplasty[2]; harvesting of pericranial duraplasty through a separate incision[3]; microdissection of cerebellar tonsils and brainstem using the operating microscope.

The patient did very well post operatively. He had complete resolution of his preoperative nausea and vomiting symptoms. His only postoperative symptoms were intermittent headaches over the pericranial graft site. These symptoms have been lessening over time since the surgery.

***Discussion***

Chiari malformations are a group of anatomic abnormalities involving the cerebellum, brainstem and craniocervical junction. They are subdivided into four types based upon specific anatomic abnormalities. Chiari malformations are often associated with spinal cord issues such as syringomyelia. In this particular case the patient had a Chiari I malformation, which includes abnormally shaped cerebellar tonsils that are displaced below the foramen magnum[1]. In adults, cerebellar tonsils that are 5 mm or more below the foramen magnum are considered indicative of a Chiari malformation. Although some studies show that the frequency of associated spinal cord cavitations is approximately 40%, other studies indicate the number may be as high as 75%[2,3]. Hydrocephalus is a much less commonly seen phenomenon with Chiari I malformations, and is noted in about 10% of cases[4]. The overall prevalence of Chiari I malformations is not entirely clear. However, over the past 30 years the prevalence seems to range between 0.1% and 0.5%[5].

In most cases, patients with Chiari I malformations do not manifest symptoms until adolescence or early adulthood[3]. Often times patients may also be asymptomatic and the diagnosis is made by incidental discovery on imaging studies[6]. The etiology of

clinical manifestations can be broken down into a few main categories: pain, syringomyelia, elevated intracranial pressure, cranial neuropathies, brainstem compression, myelopathy, cerebellar dysfunction[1,7]. Pain is the most common symptom, and may be intermittent or constant. It also tends to be worsened by physical activity, sneezing, laughing, or coughing[1,8]. Other symptoms that can be seen with Chiari I malformations are nystagmus, tongue atrophy, vocal cord paralysis, hoarseness, recurrent aspiration, and dysarthria. Patients may also exhibit symptoms of weakness, ataxia, nystagmus, nausea, vomiting, and hiccups[8].

The hallmark of diagnosis is neuroimaging, and MRI is the superior modality for diagnosing this condition[9]. In situations where a patient cannot undergo MRI, high resolution CT with sagittal reconstructions is the alternative of choice[10].

The options for management depend upon the type of malformation as well as associated neurologic symptoms. There are some who suggest that patients with Chiari I malformations without syringomyelia and without symptoms, can be managed conservatively with surveillance imaging[11]. However, there are others who suggest that these patients undergo prophylactic surgery[12]. Certainly any symptomatic patient with severe neck pain, headaches, myelopathy, cerebellar symptoms, syringomyelia, or lower cranial nerve palsies should undergo decompressive surgery.

The surgical procedures have the goal of decompression of the craniocervical junction and restoration of normal cerebrospinal fluid flow in the foramen magnum. This is usually achieved by posterior decompression with or without duraplasty. Alternatives to this include anterior decompression or a shunting procedure.

In conclusion this case represents a good example of a patient with very common symptoms of nausea and vomiting without an initial obvious diagnosis. It illustrates the importance of considering an underlying neurologic cause of these symptoms in the appropriate patient. In this case, the diagnosis of Chiari I malformation with syringomyelia, ultimately lead to surgical decompression and complete resolution of the patient's chronic nausea and vomiting symptoms.

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