

## Chiari Type 1 malformation: A case report

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### ABSTRACT

Type I Chiari malformation (CM) is an acquired or congenital disorder characterized by herniation of the cerebellar tonsils through the foramen magnum and into the spinal canal. The resulting anatomic abnormality impairs the flow of cerebral spinal fluid and results in the Chiari symptoms and signs. The purpose of this study was to report a case of successfully treated CM Type 1 that was discovered accidentally during childhood. A 9-year-old female presented to the emergency department complaining of dizziness and headache. Before her admission, she lost balance and collapsed to the ground fainting. A magnetic resonance imaging (MRI) scan of the brain disclosed herniation of cerebellar tonsils through the foramen magnum at 13 mm below the spinal canal. It is very difficult to determine the true frequency of this disorder early in childhood. A definitive diagnosis is generally made after a MRI, where the abnormal protrusion of the cerebellum toward the spinal cord can be seen.

**Key words:** Case report, Cerebellum, Chiari, Congenital malformation

Chiari malformations (CMs) are a constellation of congenital anomalies related to structural defects in the base of the skull and cerebellum, the part of the brain that controls balance [1]. There are four different categories of CMs with different degrees of severity, being CM Type I the less severe characterized by cerebellar herniation of  $>5$  mm in the upper spinal canal, rather than the posterior fossa [2]. CM is still listed as a rare disease by the Office of Rare Diseases of the National Institutes of Health. However, increased use of imaging tests has led to more frequent diagnoses [3] with an estimated prevalence of 1% in the pediatric population [4,5]. CM Type I is the most common and has been estimated to occur in one in 1000 births with a slight female predominance of 1.3:1 [6]. Possibly, the epidemiological data are underestimated since some individuals may not show any symptoms and often go unrecognized until adolescence or adulthood. Symptoms may change for some individuals, depending on the compression of the tissue and nerves and on the build-up of cerebrospinal fluid (CSF) pressure. The most common symptom associated with a CM is occipital headaches that are felt near the base of the skull and may radiate to cause pain in the neck and shoulders. The pain can be severe and may be described as sharp, brief, throbbing, or pulsating. Occipital headaches can be brought on or worsened by coughing, straining, or sneezing [2]. Additional symptoms associated with a CM may

include poor coordination and balance problems, muscle weakness, dysphagia or dysarthria, palpitations, fainting episodes, and tingling or burning sensations in the fingers, toes, or lips. Sleep disorders, especially sleep apnea and chronic fatigue, have also been described in individuals with CM.

### CASE REPORT

A 9-year-old Iraqi female presented to the emergency department complaining of nausea, vomiting, dizziness, and a 2-day history of headaches. Immediately before her admission, she lost balance and collapsed to the ground fainting. She was conscious and oriented. Medical history revealed previous attacks of brief, tension-like occipital pain that radiate to the neck and shoulders after coughing or sneezing. There was also a history of unsteady gait and loss of balance from time to time.

On arrival at the hospital, her vital signs were as follows: Body temperature, 37.8°C; pulse rate, 93 beats/min; respiratory rate, 20 breaths/min; and blood pressure, 110/70 mmHg. Physical examination revealed mild tenderness in the left temporal region, no neck stiffness, and equal bilateral pupil reaction, symmetrical tone and sensation in both upper and lower limbs. She had downbeat nystagmus, dysmetria, and ataxia, especially on the left side.

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#### Quick Response code



Plain radiographs showed no abnormalities. A computed tomography scan of the brain was performed that revealed crowding of cerebellar elements at the foramen magnum. Further work-up with brain magnetic resonance imaging (MRI) disclosed herniation of cerebellar tonsils through the foramen magnum at 13 mm below the spinal canal (Fig. 1).

The case was diagnosed as Chiari Type 1 malformation; subsequently, surgical decompression was performed with suboccipital craniotomy, C1 decompression, and duraplasty. The surgeon placed the head in a skull fixation device to hold it in place during the surgery. A strip of hair was shaved along the area of the planned incision and the scalp is prepared with an antiseptic solution. A small section of bone in the back of the skull was removed to widen the foramen magnum and create space for the brain to relieve pressure. The dura overlying the herniated tonsils was opened and a patch was sewn to expand the space. The patient had no underlying medical conditions that could lead to an immunocompromised state. She was discharged 4 days later while receiving analgesics and muscle relaxants.

## DISCUSSION

To the best of our knowledge, this is the first report of a case of Chiari Type 1 malformation in a governmental hospital in Ras al Khaimah, UAE. It is caused by a structural defect in the base of the skull and downward displacement by 13 mm, of the cerebellar tonsils beneath the foramen magnum into the cervical spinal canal in a female child. There is evidence that CM Type 1 runs in some families, which suggests having a genetic cause [6,7]. It is hypothesized that Chiari Type 1 originates as a disorder of para-axial mesoderm, which subsequently results in the formation of a small posterior fossa. The development of the cerebellum within this small compartment results in overcrowding of the posterior fossa, herniation of the cerebellar tonsils, and impaction of the foramen magnum [8]. Less often, the malformation can happen later in life from trauma, infection, or other medical problems. Anomalies of the base of the skull and spine are seen in 30–50% of patients with Chiari I malformation [9].

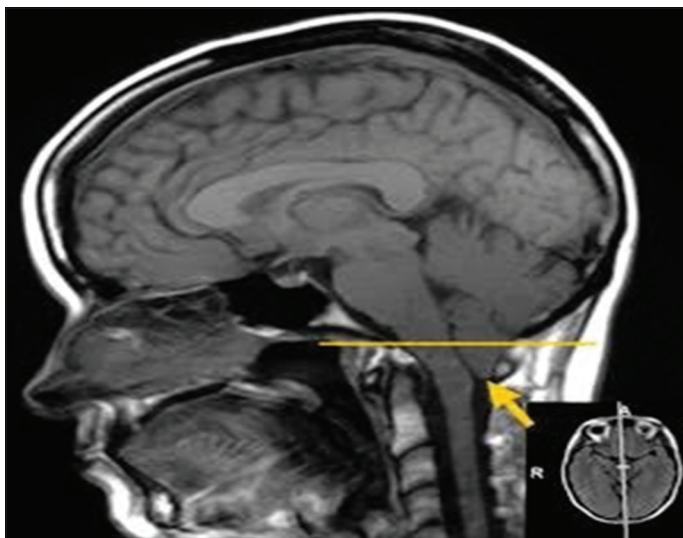


Figure 1: Sagittal view of magnetic resonance imaging of the brain

Symptoms of Chiari I develop because of the pathophysiological consequences of the disordered anatomy. Compression of the cerebellum may result in ataxia, dysmetria, nystagmus, and disequilibrium. Furthermore, disruption of CSF flow through the foramen magnum probably accounts for the most common symptom, headache, and neck pain. These symptoms are usually asymmetrical and have a tendency to develop in the side more significantly affected by tonsillar ectopia [10]. This explains the left-sided symptoms and signs of the current case report. Radiographic presence of tonsillar herniation correlated with appropriate clinical signs and symptoms before surgical intervention was undertaken. Indeed, cerebellar symptoms and occipital headaches make the decision in favor of surgery straightforward.

Conservative treatment is considered for patients with Chiari I malformations who have minimal or equivocal symptoms without syringomyelia. Nevertheless, Langridge in 2017 reported that approximately 93% of asymptomatic patients with Chiari I remained asymptomatic, even if syringomyelia was present [11]. Despite significant variance in clinical manifestations among patients, early surgical intervention is associated with better outcomes in cases of symptomatic Chiari I malformation. It has been found that the rate of decompression surgery increased in patients younger than 20 years where 53% of children were female [12]. The aims of surgical treatment are decompression of the cervicomedullary junction and restoration of normal CSF flow in the region of the foramen magnum. Preparation for surgery for Chiari I decompression was the same as for any elective surgery and depends on the patient's general health. Post-operative, the patient was carefully observed during the first 24 h in ICU for any signs of brainstem dysfunction, particularly apnea.

In our case, she was discharged home when she was ambulatory and able to eat without vomiting by the end of post-operative day 4. Incisional pain and muscle spasms were controlled with opiate analgesics and muscle relaxants. An appointment with neurology for follow-up was scheduled for her on a weekly basis for 1 month.

## CONCLUSION

Infants who are born with this condition may never develop symptoms or show symptoms, only in early adolescence or adulthood. Therefore, work-up for this entity should be guided by a thorough history and MRI, which are mandatory for the malformation to be diagnosed. Early recognition of symptoms of brainstem compression and subsequent surgical decompression can relieve symptoms and restore quality of life among children with Chiari Type I malformation.

## AUTHORS' CONTRIBUTIONS

All authors contributed to the study conception and design. Material preparation, data collection, and analysis were performed by Rasha Aziz Attia Salama, Huda Abdel Nabi, and Nihal Amir Alfons. The first draft of the manuscript was written by Rasha Aziz Attia Salama and all authors commented on previous

versions of the manuscript. All authors read and approved the final manuscript.

## ETHICAL STATEMENTS

The study was approved by the research and ethics Committee of Ras Al Khaimah Medical and Health Science University (No. RAKMHSU-REC-42-UG-M.). Written informed consent was obtained from the guardian of the patient to participate in the study.

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