CASE REPORT

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A Case of Arnold Chiari Malformation Type-1 Diagnosed Incidentally After Head Trauma

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ABSTRACT Arnold-Chiari Malformation is characterized by herniation of the tonsils of the cerebellum through the foramen magnum into the cervical spinal canal. The etiology of Arnold-Chiari Malformation remains unclear. Malformation may be accompanied by intracranial or extracranial developmental defects such as hydrocephalus, syringomyelia, encephalocele. Different classifications are made according to the accompanying malformation. Arnold-Chiari Malformation can remain asymptomatic during childhood or manifest with neurological symptoms such as headaches. Since there are many different complaints and findings in this malformation, patients present to various medical units. We present a patient who presented to the emergency department with recurrent vomiting after head trauma and was found to have Arnold-Chiari Malformation Type-1 incidentally.

Keywords: Trauma; child; vomiting; chiari; malformation

Arnold-Chiari malformation is characterized by herniation of the tonsils of the cerebellum through the foramen magnum into the cervical spinal canal.[1] The malformation may be accompanied by intracranial or extracranial developmental defects such as hydrocephalus, syringomyelia, and encephalocele.[2] The etiology of Arnold-Chiari malformation (ACM) remains unclear.[1,3]

Data on the incidence of Arnold-Chiari malformation are limited because it is often asymptomatic in childhood. However, according to radiologic imaging studies, the prevalence is thought to be around 1%.[1]

The most common complaint of patients with ACM is pain. Head, neck, shoulder, and back pains are common, characterized by Valsalva maneuver or coughing, and radiate from the occipital region to the frontal region.[4] Other symptoms include weakness, spasticity, and ataxia as a result of brain stem and spinal cord dysfunction or upper/lower motor neuron dysfunction due to direct compression. Sensory deficits, respiratory dysfunction, and lower cranial nerve deficits such as dysarthria, dysphagia, soft palate weakness, decreased pharyngeal reflex, and vocal cord paralysis. Additionally, it has also been reported that scoliosis development can occur in cases with hydrocephalus.[4,5]

ACM is clinically classified according to the degree of herniation and the presence of other accompanying anomalies.[1-3] Clinical findings and prognosis of the patients vary according to the degree of herniation and accompanying anomalies. Patients may be asymptomatic in childhood or surgical operation may be indicated.[1,2]

In this study, we report a patient who presented to the emergency department with recurrent vomiting after head trauma and was found to have ACM Type-1 incidentally.

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CASE REPORT

A male patient who was 8-year-old was presented to the emergency department with recurrent vomiting. It was learned that his vomiting consisted of stomach contents. He vomited 7 times on the same day, his stool was normal, and he had no other complaints. When the medical history is detailed, it is learned that the patient fell from a wall 2 meters high to the hard ground 4 days ago; he did not have a headache or vomiting after the fall. Cranial computed tomography (CT) was performed on the same day and was normal.

In the patient's initial evaluation, blood pressure was 90/60 mmHg, peak heart rate was 100/min, respiratory rate was 24/min, Spo2: 98%, and body temperature were 36.4 °C.

On abdominal examination was comfortable, and no defiance rebound tenderness was detected. The Glasgow Coma Scale was 15 on neurological examination, light reflex was obtained bilaterally, and no lateralization sign and nuchal rigidity were detected. Other system examinations were evaluated as normal.

Hemoglobin in the blood test of the patient: 12.3 g/dL leukocyte: 7,920/uL, platelet: 240,000 e3/uL blood glucose level was 95 mg/dL, creatinine 0.35 mg/dL, uric acid 3.4 mg/dL, sodium: 139 mEq/L potassium: 4.29 mEq/L aspartate aminotransferase 29 IU/L alanine aminotransferase 12 IU/L C-reactive protein: 3.8 mg/L and urinalysis was normal. No acidosis was detected in the blood gas.

Since the patient had recurrent vomiting and a history of head trauma, a cranial CT scan was performed and found to be normal. No papillary edema was found on ophthalmologic examination. Ear, nose, and throat examination for the etiology of central vomiting was normal.

The patient was admitted to the service for follow-up. A brain magnetic resonance imaging was performed. Brain MRI revealed that the cerebellar tonsil extended inferiorly from the foramen magnum and was compatible with ACM Type-1 (Figure 1).

A wide range of clinical manifestations characterizes ACM, and the patient's recurrent vomiting was associated with ACM. The patient was followed up by neurosurgery.

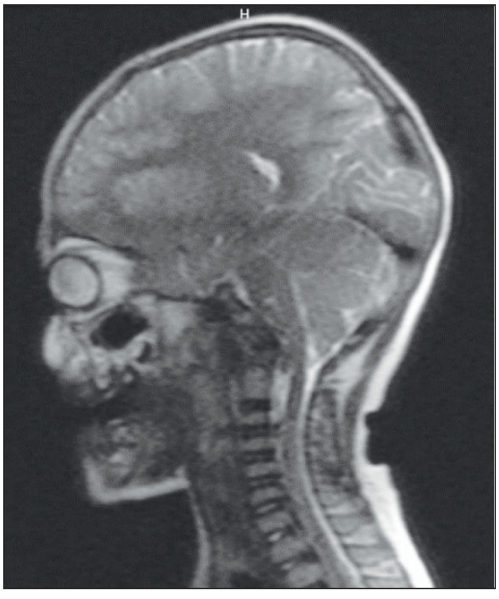

FIGURE 1: Herniation in cerebellar tonsils.

Written consent was obtained from the patient's parents for the case presentation.

DISCUSSION

The etiology of ACM remains uncertain.[1,2] There are several theories to explain its etiology. Among these theories are hydrodynamic, overgrowth, stretching traction, neuroschisis (dysraphic), arrested development, small posterior fossa, and primary mesodermal insufficiency.[2,3] The most widely accepted theory is the mesodermal defect in the craniovertebral junction.[2,3]

Clinically, ACM Type I may be asymptomatic or present in late childhood or adulthood with headaches and focal neurologic signs characterized by coughing. There are different types of ACM.[3,4] Types II, III, and IV are congenital and clinically significant. The nosographic ranking of type 0 or 0.5 Chiari malformation or "Chiari-like" (symptoms without tonsillar herniation), Type 1.5 ACM (intermediate between Types I and II), and complex Chiari is controversial and not universally accepted.[1]

Since there are many different complaints and findings in this malformation, patients present to various medical units.[3-5]

Grahovac et al. retrospectively evaluated 16 operated cases with ACM Type-1. In this study, it was reported that 12 patients presented with headache symptoms, including irritability, inconsolable crying, restlessness, and head shaking backward; 10 patients presented with oropharyngeal and/or respiratory symptoms, including vomiting, retching, snoring, sleep apnea, respiratory arrest and/or vocal cord paralysis, and only one patient had segmental cervical hydromyelia.[6]

Our patient presented to the emergency department with vomiting which is one of the rare findings of ACM. When our patient was evaluated with differential diagnoses for the etiology of vomiting, it was not compatible with gastrointestinal pathologies. Following MRI, which was thought to be associated with central pathologies, ACM was detected and her vomiting was associated with this condition.

Vomiting is a complicated response mediated by the emetic centre located in the lateral reticular formation of the medulla.[7] This center receives input from various areas of the body which include vagal afferents from the gastrointestinal tract, psychogenic stimuli from the cerebral cortex and stimuli from the vestibular and visual areas as well as the chemoreceptor trigger zone.[7,8] In ACM, vomiting may be observed with stimulation of the vomiting center as a result of direct compression of the brain stem or in case of increased intracranial pressure.[8] The pathogenesis of vomiting in our patient was associated with direct compression of the brain stem due to herniation after exclusion of other acute pathologies.

Patients with asymptomatic ACM are followed up medically. Nonsteroidal anti-inflammatory drugs and muscle relaxants are used in medical treatment for pain. Surgical treatment is indicated in patients with persistent clinical symptoms or progressive symptoms during follow-up and especially in patients with neurologic deficits.[5,6,9]

The patient was incidentally diagnosed with ACM following recurrent vomiting post-head trauma. Vomiting in children at the emergency department is common, attributed to gastrointestinal or less commonly, central nervous system issues. Tailoring examinations to the medical history is crucial, as in our case where cranial imaging was indicated for suspected central causes of vomiting. This case highlights the rarity of vomiting in ACM patients.

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Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

This study is entirely author's own work and no other author contribution.

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