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Case Report on Chiari Syndrome

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Chiari Malformation is a rare condition. A condition known as Chiari malformation occurs when brain tissue spreads into the spinal canal. When a portion of your skull is excessively small or malformed, it presses on your brain and forces it downward. Chiari malformation is a rare occurrence, although the increased use of imaging testing has resulted in more diagnosis.

Case Presentation: A 18-year-old boy was admitted to the hospital with the following symptoms: Tingling sensation, numbness over left hand since 2 to 3 months. Neck bend toward right side, pain in left hand since 6 month. Difficulty during eating by hand since 2 to 3 month. On physical examination, indicated a bright attentive person with pale conjunctiva and no symptoms of icterus. He had a tachycardia, bilateral pitting pedal edema and a swollen abdomen with shifting dullness, all of which pointed to as cites. He had a history of intermittent abdominal pain. On admission he complaint of new onset of dyspnea on exertion, fatigue and abdominal swelling. The rest of all physical examination was normal, with no skin changes and an intact arterial pulses in all four extremities.

Conclusion: The primary focus of this case study is on professional management and outstanding nursing care, which may provide the holistic care that Chiari Syndrome necessitates while also effectively managing the challenging case. After a full recovery, the patient's comprehensive health care team collaborates to help the patient regain his or her previous level of independence and satisfaction.

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Keywords: Chiari syndrome; tingling sensation; swollen abdomen; numbness.

1. INTRODUCTION

Chiari syndrome is characterized by a clinical triad of abdominal discomfort, ascites, and hepatomegaly due to a hepatic venous outflow obstruction [1,2]. Although it is recognized as a cause of Chiari syndrome, it is extremely rare, with just a few cases reported [3]. We provide a unique case of Chiari syndrome in a patient who developed hepatic vein and Inferior vena cava thrombosis at the same time, leading to the development of cirrhosis, and exhibited significant improvement with treatment [2,4] Chiari syndrome with severe vascular consequences is considerably more common in young adult male patients; we present a rare case of Chiari syndrome [5].

The general population's prevalence is believed to be somewhat less than one in 1000 [6,7]. The majority of these patients show no signs or symptoms. Patients who have had diagnostic imaging for unrelated reasons are frequently found to have Chiari malformations [8,9]. Type I Chiari malformation develops as the skull and brain mature [10]. As a result, symptoms and indicators may not appear until later in childhood or adulthood [11]. Chiari malformation type II and type III are present at birth in children (congenital) [5,12] Chairi syndrome is a rare disease [13]. The form, severity, and related symptoms all influence how Chiari malformation is treated8.Treatment options include regular monitoring, medicines, and surgery [14,15].

When a portion of the skull is smaller or malformed, it presses on the brain and forces the cerebellum down into the spinal canal, causing Chiari malformations [16]. Pressure on the cerebellum and brain stem may disrupt functions controlled by these organs and obstruct the flow of cerebrospinal fluid (CSF), a clear liquid that surrounds and cushions the brain and spinal cord [1,17]. When a portion of the skull is smaller or malformed, it presses on the brain and forces the cerebellum down into the spinal canal, causing Chiari malformations [16,18]. Pressure on the cerebellum and brain stem may disrupt functions controlled by these organs and obstruct the flow of cerebrospinal fluid (CSF), a clear liquid that surrounds and cushions the brain and spinal cord [13]. This is the deformity's most severe and uncommon variant [3]. The cerebellum isn't developing properly [2]. Other abnormalities of the brain and brainstem may be present [8,12].

The cause of a type I congenital Chiari malformation is unknown [6,13]. The deformity could be caused by a malfunction during embryonic development. It's possible that it's caused by coming into contact with dangerous substances when pregnant [1,2]. It could also be linked to hereditary issues that run in families. After birth, a person develops an acquired Chiari malformation type I. Excess spinal fluid leakage from the lower back (lumbar) or chest (thoracic) portions of the spine causes it [5,4]. This can occur as a result of an accident, exposure to toxic substances, or illness [2,19].

2. PATIENT INFORMATION

A 18- year -old boy was admitted to the Acharya Vinobha Bhave Rural Hospital with complaints of tingling sensation. numbness over left hand since 2 to 3 months. Neck bend toward right side, pain in left hand since 6 month. Sleep apnea, Difficulty during eating by hand since 2 to 3 month. On physical examination, indicated a bright attentive person with pale conjunctiva and no symptoms of icterus. He had a tachycardia, bilateral pitting pedal edema and a swollen abdomen with shifting dullness, all of which pointed to ascites. He had a history of intermittent abdominal pain. On admission he complaint of new onset of dyspnea on exertion, fatigue and abdominal swelling. The rest of all physical examination was normal, with no skin changes and an intact arterial pulses in all four extremities. He has no any family history of this disease. The patient condition agitated. He couldn't keep up with his hygiene. The patient family is from a working class family. Both communicable and non communicable disease were absent in his family. with relative, neighbours, and other family members, he and his family had good interpersonal relationships when he was admitted, RBS test ,Magnetic Resonance Imaging (MRI),and Computerized Tomography Scan (CT Scan), Administration of analgesic and antibiotics as per physician orders.

3. PHYSICAL EXAMINATION

Chiari malformation patients frequently appear normal. Patients with Chiari malformation frequently have decreased coordination, sensory/motor deficits, irregular gait, nystagmus, scoliosis, and autonomic dysfunction, weakness on physical examination. so treatment was started as soon as possible.

4. DIAGNOSTIC ASSESSMENT

Blood test: Haemoglobin% -14.5%, total Red Blood Cell count-5.43 million/cu.mm, total White Blood Cell count-5900/cu.mm, total platelet count-2.28lacs/cu.mm, In patients with chiari syndrome High protein concentrations (>2 g/dL) are common in patients but this may not be the case in those with the acute type. In most cases, the white blood cell (WBC) count is less than 500/L. In most cases, the serum ascites–albumin gradient is less than 1.1. (except in the acute forms of the disease) faecal culture for enteric pathogens including *Clostridium difficile* toxin, *Giardia* antigen, *Cryptosporidium* antigen and other ova and parasites. The elevation of his liver enzymes remained unexplained, as he had no history of alcohol abuse and serological studies for hepatotropic viruses were negative. Laboratory studies performed to evaluate for Wilson's disease, alpha-1 antitrypsin deficiency, autoimmune hepatitis, non-alcoholic steatohepatitis and primary biliary cholangitis were also negative.

Medical management: The severity and characteristics of Chiari malformation will determine how you are treated. If you have no symptoms, your doctor would most likely recommend no therapy other than regular checkups and MRI to keep an eye on you. Your doctor may prescribe pain medication if headaches or other types of discomfort are the major complaint. Patients with Chiari I anomalies who do not have syringomyelia and have mild or ambiguous symptoms can be treated conservatively. Analgesics, muscle relaxants, and the occasional use of a soft collar can be used to alleviate mild neck pain and headaches. Surgical treatment should be offered to patients who are symptomatic. A thorough analysis of the literature found symptomatic patients who did not have surgery.

Nursing management: Chairi syndrome is treated with a combination of lifestyle changes, medication and if necessary, special procedures, therapy or surgeries. Nurses will determine which treatment is best for the patient condition. The patient vital signs are meticulously documented. To help the patient with chiari syndrome, the nurse will need to work diligently. Take proper medication, improve balance problems, improve swallowing problems, managing sleep apnea, healthy diet and blood level. Assist in symptomatic treatment. Manage tingling sensation, numbness and eating difficulty. According to patient family members, excellent nursing care was provided. Interact to improve the patient condition and reduce risk of complications.

5. DISCUSSION

Chiari Syndrome is defined as any pathologic event that causes the normal blood flow out of the liver to be interrupted or reduced, either inside the hepatic veins or the Inferior vena cava [20,18]. Portal vein thrombosis can occur when the extrahepatic venous system is occluded [21,10] Venous obstruction in the liver occurs in the following proportions: 62 percent in the hepatic veins, 7% in the Inferior vena cava, 31% including both, and 14% in the portal vein [22]. Ascites, stomach pain, hepatomegaly, and, in certain cases, hepatic necrosis leading to severe liver failure are common symptoms of Chiari Syndrome [14,23]. Hepatic congestion, portal hypertension, and ascitis are all symptoms of Chiari Syndrome [11,24].

The cerebellar tonsils are displaced downward by more than four millimeters beneath the foramen magnum into the cervical spinal canal during fetal development, resulting in this deformity [7,25]. The regular pulsations of CSF between the spinal canal and the intracranial space may be blocked by this displacement [15,16]. This type of Chiari malform The medulla, fourth ventricle, and cerebellum are displaced downward into the cervical spinal canal, and the pons and fourth ventricle are elongated in this abnormality [22,25]. This form is virtually exclusively seen in myelomeningocele patients [6,10]. Myelomeningocele is a congenital disorder in which the spinal cord and column do not close properly during fetal development, resulting in a condition known as myelomeningocele.ation is linked to syringomyelia/hydromyelitis [27,26]. A piece of the cerebellum and/or brainstem pushes out through a defect at the back of the head or neck in this deformity, which is called dysraphism [25,20]. These abnormalities are extremely rare, and those who survive have a high rate of early mortality or severe neurological impairments. If therapy is decided upon, the defect must be closed as soon as possible [7,22]. This is the most severe and rare variant of the deformity [25,28]. The cerebellum does not grow properly. Other brain and brainstem abnormalities may be present [11,15]. The majority of newborns born with this deformity do not live to see their first birthday [20,28].

6. CONCLUSION

The cerebellum is a part of the brain that affects movement coordination and is generally found in the posterior fossa of the skull. The cerebellum is usually divided into two halves, or hemispheres, with a narrow middle component known as the vermis between them. The tonsils are two tiny protrusions that run along the underside of each hemisphere's surface. The fourth ventricle is a region in front of the cerebellum that is filled with cerebrospinal fluid (CSF) (and behind the brainstem). All of these structures sit right above the foramen magnum, which is the main aperture at the base of the skull through which the spinal cord enters and links to the brainstem. The general population's prevalence is believed to be somewhat less than one in 1000. The majority of these patients show no signs or symptoms. Patients who have had diagnostic imaging for unrelated reasons are frequently found to have Chiari malformations.

Chiari malformations occur when a part of the skull is smaller or deformed, pressing on the brain and forcing the cerebellum down into the spinal canal. Pressure on the cerebellum and brain stem can impair functions and block the flow of cerebrospinal fluid (CSF), a clear liquid that surrounds and cushions the brain and spinal cord. When a portion of the skull is smaller or malformed, it presses on the brain and forces the cerebellum down into the spinal canal, causing Chiari malformations. Pressure on the cerebellum and brain stem may disrupt functions controlled by these organs and obstruct the flow of cerebrospinal fluid (CSF), a clear liquid that surrounds and cushions the brain and spinal cord.

CONSENT

While preparing a case report and for publication patients informed consent has been taken.

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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