Case Report

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Arnold Chiari Malformation Type 1 with Cerebellar Tonsil Herniation through Foramen Magnum: A Case Report

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Abstract

Arnold Chiari malformation is an uncommon disorder that mainly occurs in the posterior fossa. The prevalence is relatively unknown, but it is estimated that around one child will suffer from Arnold Chiari malformation in every 1000 births. In this case study, we presented a case of a 45-year-old female suffering from Arnold Chiari malformation type I. Following her clinical examination, she underwent foramen magnum decompression surgery.

Keywords: Arnold-Chiari malformation, case report, epidural anaesthesia, tonsillar herniation

Introduction

Arnold Chiari malformation is a structural abnormality of the brain's cerebellum, which controls stability (1). In this syndrome, there could be an herniation of the cerebellar tonsil into the spinal canal through the foramen magnum to a complete absence of the cerebellum. Depending upon the severity, the malformation is divided into four types. Which are Arnold Chiari malformations type I, type II, type III, and type IV (2). Previously, scientists held the belief that the disease was extremely rare. But due to the advancement of medical imaging techniques such as CT scans and MRIs, we could see the disease is not rare anymore (1). However, the majority of the type I CM are asymptomatic, and not all the patients can afford to access the imaging techniques. Therefore, it is believed that the worldwide cases are under estimated. Some of the risk factors are increasing age, increasing size of cerebellar tonsils greater than 5 mm, being female, ethnicity, and race. To make a diagnosis, further causes of cerebellar tonsillar ectopy must be ruled out in addition to a physical examination, additional testing, neurological assessment, and neuroimaging procedures including CT and MRI (3). In India, we don't have any specific data on the total number of people who got affected by the ACM type I, as it is usually asymptomatic until adulthood (4). Moreover, the true pathogenesis of the disease is still unknown, so to avoid further challenges, it is crucial to diagnose the illness as soon as possible. In this study, we report a case of a 45-year-old female who presented with a chief complaint of suboccipital headache and paraesthesia of limbs.

Case Report

A 45-year-old female was admitted to Apollo Hospital in the department of neurosurgery on 28 June 2024 with the complaint of headache and neck pain, where she was provisionally diagnosed with an Arnold Chiari malformation. Later after the MRI, her final diagnosis was an Arnold Chiari malformation with cerebellar tonsil herniation through foramen magnum.

Clinical Signs

On examination, patients complain of headache and neck pain since three months, unable to sit for long durations. However, the patient did not have any signs of tingling sensations or numbness of limbs. CVS: S1 S2 positive, P/A soft and nontender, PAC panel II, HMF: intact, CN: intact. Furthermore, the patient did not have a history of falls or previous episodes of vomiting.

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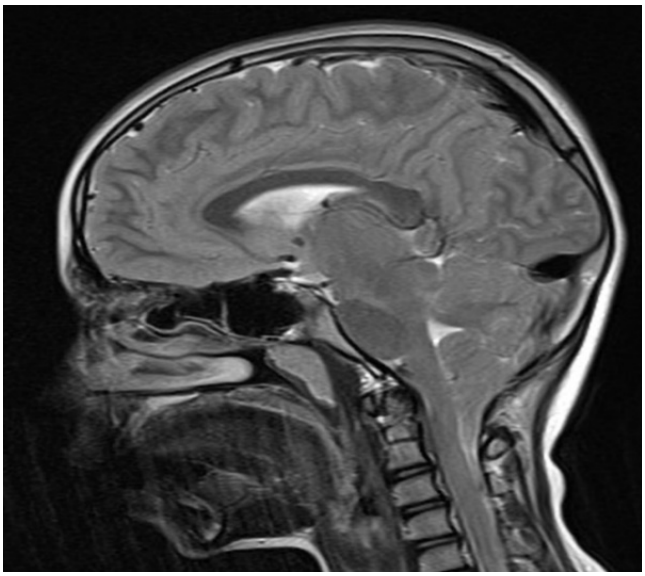

Figure 1. MRI shows tonsillar herniation

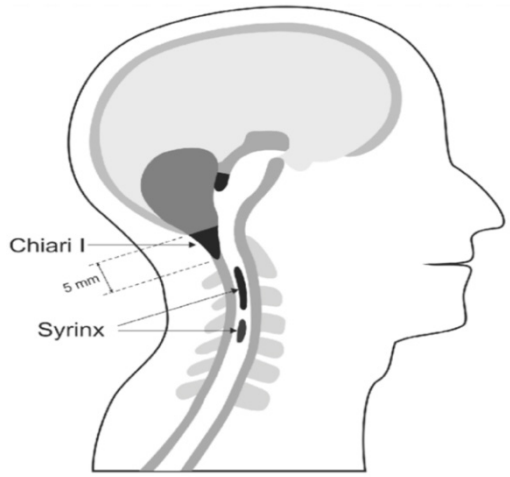

Figure 2. Anatomical positioning of the chiari 1 and syrinx

Investigations

On the arrival of the patient, the relevant investigations were done. Blood grouping and typing test results show ABO group: B, Rh (D) type: positive, Anti-HCV antibodies: non-reactive; HBsAg: non-reactive; retrovirus test: non-reactive. EDTA result shows platelet count: 163 103 / mm3, WBC count: 7.63 103 / mm3, neutrophils: 71.0*%, lymphocytes: 23.9*%, eosinophils: 2.0%, monocytes: 2.6%, basophils: 0.5%, ESR: 15 mm/1st hr, Hb: 11.5* g/dl. LFT function test reveals albumin serum: 3.4* g/dl, globulin serum: 3.7* g/dl, albumin-globulin serum: 1*. MRI cervical spine reveals disc osteophyte complex at C5-C6 disc level intending the ventral thecal sac without spinal canal stenosis or significant nerve compression and low lying cerebellar tonsils. It is noted that the tip of the cerebellar tonsil is approximately 2.9 mm below the level of the foramen magnum.

Therapeutic Intervention

After the confirmation of Arnold Chiari malformation type, I foramen magnum decompression surgery was done, and all the vitals were stable post-operatively. Furthermore, the patient was prescribed Tab peuron-CD3 10' S OD X 3 months, Cap sutril Q10 OD X 6 weeks, Tab pregadoc NT OD X HS X 3 months, Inj ceftriaxone 1gm X 1V X 12 hourly, Inj Pan 40 mg X IV X 24 hourly, Injdexa X 4 mg X IV X 8 hourly, and Inj MVI 100 mg with 100 ml NS X IV X 24 hourly. Furthermore, the patient was also advised to use a cervical collar.

Discussion

Arnold Chiari malformation is commonly diagnosed with the help of MRI testing. However, the fundamental challenge lies in the early radiological diagnosis. Still, the exact cause is unknown, but it was suggested that MTHFD 1 G1958A gene polymorphism is strongly linked with the neural tube defect. Furthermore, few more genes are detected that are associated with the adult Chiari malformation type 1, such as ALDH1A2, CDX1, and FLT1 (5). The herniation is diagnosed if one or both cerebellar tonsils are 5 mm or below the basion-opisthion line as measured on the midline sagittal T1-weighted MRI scan. However, the pathophysiology is far more complex, where there could be different tonsiller descent. Several previous studies also reported different unique and distinctive features of the tonsillar descent (6). In our case, the patient was suffering from Chiari type 1, which is most common among all the Chiari types, and it mostly affects the females, with 0.5 to 3.5% of the population. In this type, the bones of the sull base were often underdeveloped, which results in posterior fossa volume reduction (7) and is frequently associated with syringomyelia. The management of the Arnold Chiari malformation depends upon the severity and should be corelated clinically. Asymptomatic patients without having hydrocephalus or syrinx can be managed conservatively with a six-month follow-up period. However, the diagnosis should be done early to have a better prognosis. If the patients are symptomatic with symptoms like myopathy, severe headache and neck pain, medullary compression, and syringomyelia, they usually require a ventriculo-peritoneal shunt. Cranial cervical decompression surgery can also be performed in case of the presence of syrinx (8). In this case, foramen magnum decompression surgery was done, and the surgery was successful. A post-operative check was done for several days, and patient vitals and all the other parameters were stable, and there was no sign of any infection. There was optimal pain control during the surgery due to the epidural anesthesia, and the patient was able to get up after 24 hours of surgery.

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Conclusion

Arnold Chiari malformation is common nowadays. However, many times patients are undiagnosed due to the lack of symptoms. However, early diagnosis is important to manage the symptoms and also to improve the quality of life. Also, the physicians should consider the possibility that Arnold Chiari malformation in cases of acute respiratory failure is unexplainable.

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Conflicts of Interests: None declared

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