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A Rare Case of Arnold Chiari Malformation-Type 1

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ABSTRACT:

The most prevalent 'Arnold Chiari type I' malformation typically goes unnoticed until maturity. To make a diagnosis, 'the cerebellar tonsils' protrusion from the borders of the inner foramen magnum to their inferior most section is commonly measured (measurement is from the ophisthion to the basion). Tonsils are regarded as normal if they are located above the foramen magnum, benign tonsillarectopia is used to describe tonsil lengths of 5 mm, and Arnold Chiari type I malformation is used to describe tonsil lengths of >5 mm.

Keywords: Arnold Chiari 1 malformation,headache,mild intellectual disability,neural tube defects,case report.

1. Introduction

Hans Chiari, an Austrian pathologist (1851-1916), initially identified Arnold-Chiari deformities in a juvenile postmortem specimen in 1891.The condition affecting the back of the

brain has been given the name Arnold Chiari malformation in honour of his professor Dr. Arnold.[1]

The exact cause of the disorder's aetiology is yet unknown. The most prevalent 'Arnold Chiari type I' malformation typically goes unnoticed until maturity. To make a diagnosis, 'the cerebellar tonsils' protrusion from the borders of the inner foramen magnum to their inferior most section is commonly measured (measurement is from the ophisthion to the basion).[2].Tonsils are regarded as normal if they are located above the foramen magnum, benign tonsillarectopia is used to describe tonsil lengths of 5 mm, and Arnold Chiari type I malformation is used to describe tonsil lengths of >5 mm.[3].Arnold Chiari malformation is rapidly becoming more common recently due to increased detection by many diagnostic techniques and advances.

Background

A set of diverse illnesses known as 'Arnold Chiari malformations' include 'AC- I malformation', which is distinguished by a type of structural defect in cerebellum and base of cranium.

In the past, Hans Chiari identified a few baby postmortem hindbrain anomalies; they were known as Chiari malformations. It comes in four different varieties.

Chiari 1	• Downward displacement of medulla and cerebellar tonsils • Syringomyelia
Chiari 2 (Arnold-Chiari Malformation)	• Herniation of the cerebellar tonsils • Hydrocephalus • Kink in the medulla • (Myelo)meningocele • Syringomyelia
Chiari 3	• Further herniation of the cerebellum below the foramen magnum forming an encephalocele in addition to spina bifida
Chiari 4	• Hypoplasia/Aplasia of the cerebellum with spina bifida

Table 1: Represents Types of Arnold Chiari Malformation

The Arnold-Chiari I malformation, which is thought to affect 1 in 1000 infants, is the most prevalent of the four forms.The 'cerebellar tonsils' of AC type I malformation (also known as CM-I or type I ACM) herniate and move downhill beyond foramen magnum.On neuroimaging, tonsils that are five mm or more (often three mm) or more below the Magnum of foramen is consistent with an ACM.[4]

CM nature is still not fully understood. The average age at presentation for CM-I is typically between 15 and 18 years, and symptoms typically do not appear until adolescence or maturity. The following processes frequently contribute to the symptoms of type I ACM: compression of the medulla, cerebellum, and higher spinal cord; and obstruction of (CSF) passage through the 'foramen magnum'. Compression of the medulla and spinal cord may cause SC damage, lower CN dysfunction, and nuclear dysfunction. Additionally, a variety of neurological symptoms like dysmetria, dysequilibrium, nystagmus, and ataxia may be brought on by cerebellar compression.[5],[6].

Pain is most likely the symptom that is most frequently experienced when the CSF flow via the foramen magnum is disrupted.

The varying presentation may result in an incorrect diagnosis.

Case Presentation

A 13 year old adolescent male,who is healthy without a history of comorbidities came to our paediatric op with complaints of intermittent headache and giddiness on and off for past 1 year. Child born out of second degree consanguineous marriage.The patient had headache in the suboccipital region and frontal region.Attimes pain radiates forward behind his eyes. The headache intensifies on cough,laughing and bending over.child had very poor scholastic performance.child had IQ-64 with mild intellectual disability.

Figure1: Shows The Facial Dysmorphism.

Examination

His blood pressure was found to be 110/90 mmHg

On general examination child had dysmorphicfacies with wide nasal bridge with low set ears,high arched palate and hypertelorism. Child had unsteady gait with dysmetria. Other neurological examination was found to be normal.

Diagnosis

Figure 2

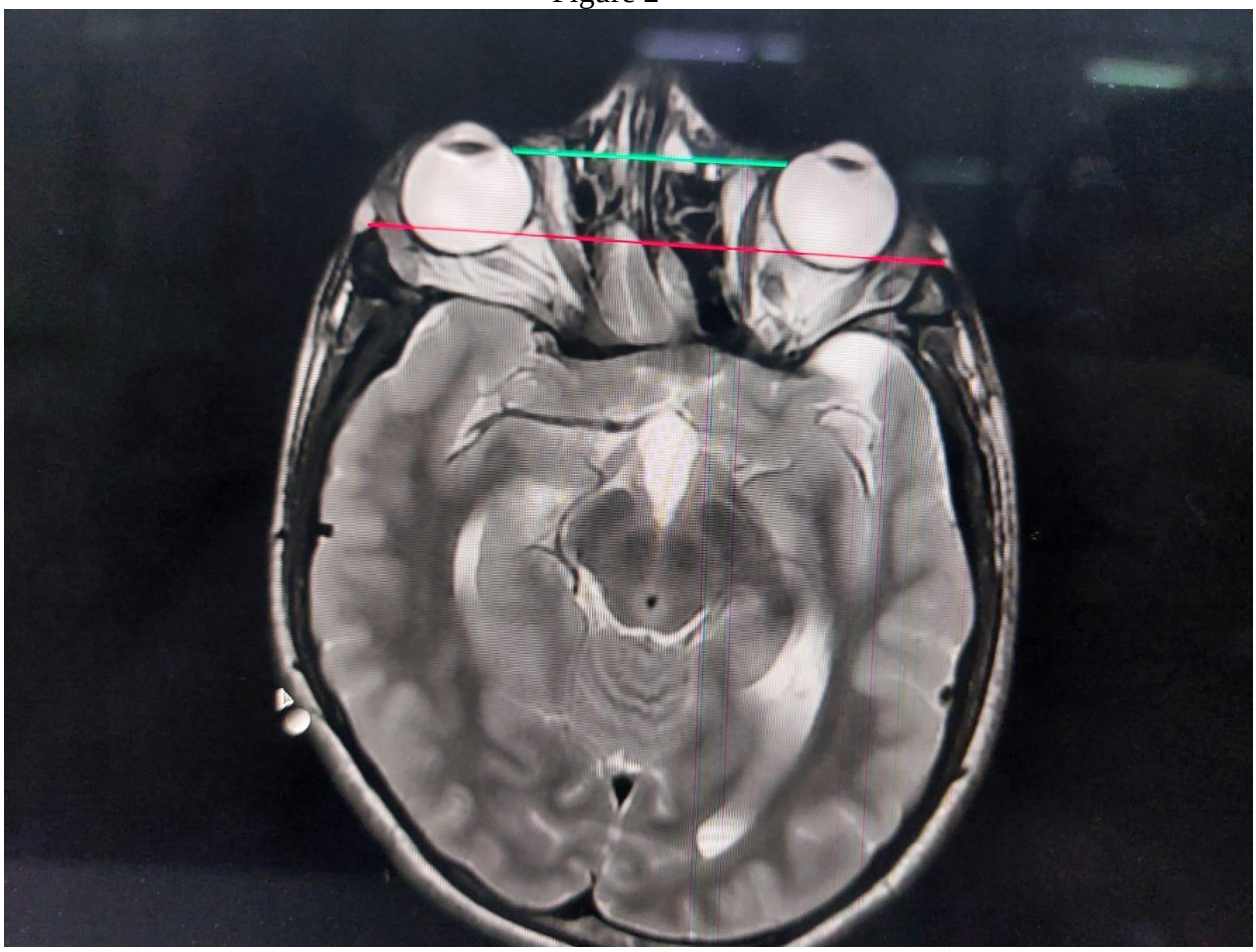
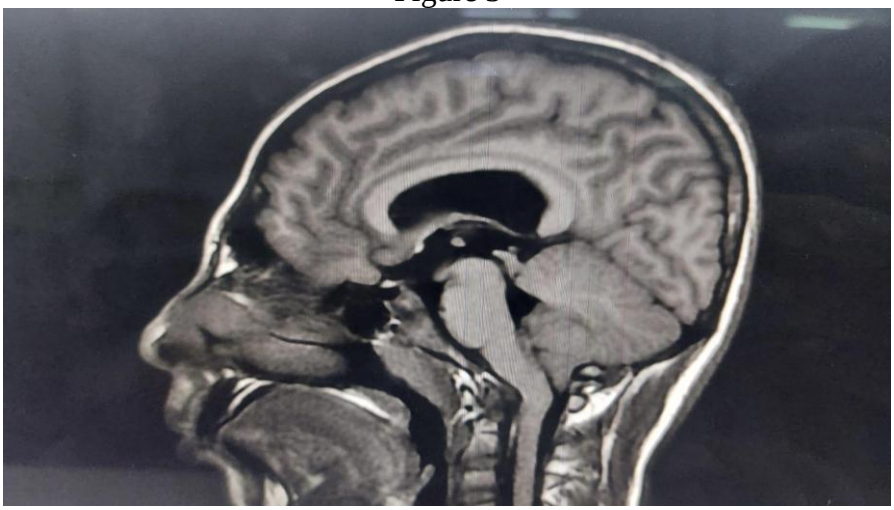


Figure 2: Shows Mri Brain

Microcephalic with asymmetry of bilateral cerebral and cerebellar hemisphere left more than right side.
Herniation of cerebellar tonsil into upper cervical canal through foramen magnum.
Prominent subarachnoid space in left temporal region.
Few subarachnoid granulations in body of sphenoid on left side.
In screening of Orbit
Tortuous left optic nerve.
Narrowed retrobulbar part of bilateral bony orbit left more than right
Hypertelorism
Bilateral optic disc pallor was seen on fundus examination.
Screening of CSF revealed
Reduced CSF flow at foramen magnum level involving both anterior and posterior aspects of cervicomedullary junction posterior more than anterior and tonsillar space.
FIGURE 3: shows CT PARANASAL SINUS
Adenoid and tomsillar hypertrophy
Deviated nasal septum to the left with septal spur impinging of left inferior turbinate
Asymmetry in ethmoid sinus roof
Aplastic right frontal sinus.

Figure 3



Treatment

Neurosurgeon opinion was sought who confirmed the diagnosis of ARNOLD CHIARI MALFORMATION TYPE I.Foramen magnum decompression surgery was planned.During surgery cerebellar tonsil was found to be completely occluding the fourth ventricle which was resected allowing normal flow of csf.

2. Discussion

The most prevalent form of ACM, Type I, is thought to affect 1 in 1000 infants and is largely asymptomatic.[7] Cases of ACMs are frequently discovered by chance while being investigated for unrelated issues.Because this categorization is based on old information, primarily from the pre-MRI era, ACM-I is no more considered to be a rare defect by (ORDR). The frequency of ACM case findings has increased with the development of MRI.[8],[9].

Males have a lower prevalence of type I - ACM, which ranges in prevalence from 0.1% to 0.5%.[10] There is strong evidence from recent studies that chromosomes 9 and According to other experts, type I ACM is a condition with main paraaxial mesodermal origins that may have an impact on the development of axial skeletal defects and several neurological malformations.[11] It can also develop in later days if the thoracic regions of spine experience significant spinal fluid drainage as a result of trauma, illness, or infection.[12] This condition is known as secondary or acquired CM.Compared to secondary CM, primary CM is substantially more frequent.[13]

A frequent structural condition known as type I - ACM is characterised by cerebellar syndrome, lower CN palsy, sensory loss, and motor deficits.[14].Neurological signs and symptoms of Type I- ACM include sensory deficit, lower CN palsies, headache,Pain in neck,ataxia.[15]

Outcome of the Case

At 8 weeks postoperative follow-up visit, child presented without headache and giddiness. Vitals found to be normal. Patient's parents were counselled that the recovery process will be ongoing and they were advised to follow up with Neurosurgeon once in every 3 months.

3. Conclusion

As a result, ACM -I is no more regarded as an uncommon condition,thus we implore clinicians to take ACM as well as other neurological reasons into consideration whenever patient presents with long term headache ruling out other diagnosis.In order to stop the progression of spinal

cord compression and subsequent neurologic damage, early detection and diagnosis by MRI are essential. Symptomatic cases might need surgical intervention.

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