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Review Article

Chiari malformations: not every disability is visible

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ABSTRACT

In Chiari malformation (CM), portions of the brain or spinal cord is pushed down through the foramen magnum, which is located at the bottom of your skull. This causes a portion of the brain to be in contact with the part of the spinal column-usually the spinal cord. The cerebrospinal fluid, however, may be pushed up through the hole making it difficult to detect in this condition. These are a collection of malformations affecting the cerebellum, pons, and medulla oblongata in the posterior fossa and hindbrain. Problems caused by these malformations range from the lack of the cerebellum to the cerebellar tonsils protruding through the foramen magnum, with or without additional intracranial or extracranial anomalies such as hydrocephalus, encephalocele, syrinx, or spinal dysraphism. CMs can be of several types and are more common in females than in males and in children than in adults. This is because they are growing and developing at a rapid rate, which leads them to have a greater amount of skull growth. In this review article, we discuss the various types of CMs, their pathophysiology, clinical features, imaging characteristics, treatment, and why they form an important complex of rare disorders necessary to be diagnosed early.

Keywords: Chiari malformation, Arnold-Chiari, Syringomyelia, Hydrocephalus, Cerebellum

INTRODUCTION

Chiari malformation (CM) is regarded as a congenital disorder, despite the fact that acquired versions of the disorder have been identified. Professor Hans Chiari, a German pathologist, first identified some malformations of the brain at the spine-skull junction in the 1890s. He divided them into categories I, II, III, and type IV based on severity. He discussed the instance of a female patient, age 17, whose tonsils had grown into cone-shaped projections that were pushed into the spinal canal along with the medulla.[1] The Chiari type II deformity was later referred to as "Arnold-Chiari malformation".

A structural abnormality in the cerebellum known as the CM is defined by the downward displacement of one or both cerebellar tonsils through the foramen magnum (the opening at the base of the skull). The posterior fossa at the base of the skull is where the cerebellum typically resides and regulates the coordination of movements. The vermis is a slender central region located between the two lateral hemispheres of the cerebellum. The tonsils are two little protrusions that run along the underside of the hemispheres. Cerebrospinal fluid (CSF)-filled chamber called the fourth ventricle is situated in front of the cerebellum (and behind the brainstem). The foramen magnum, the greatest aperture at the base of the skull through which the spinal cord enters and links to the brainstem, is directly above all of these anatomical features.

One in thousand people is thought to be affected by the condition in the general population. Most of these patients have no symptoms.[2] Patients who have had diagnostic imaging for unrelated reasons frequently have CMs discovered coincidentally. Congenital CM is three times more likely to occur in women than in men. People of

Celtic ethnicity are more likely to have type II abnormalities. Congenital CM cases may be explained by genetic and evolutionary reasons. At birth, a baby's brain weighs about 400 grams, and by age 11, it has tripled to 1100 to 1400 grams. In order to accommodate the expanding brain, the cranium triples in size from 500 cm^3 to 1500 cm^3. The skull experienced various modifications as the human species evolved to accommodate the expanding brain. The skull experienced various modifications as the human species evolved to accommodate the expanding brain. The evolution brought about changes in the skull's size and shape as well as in the basal angle and basicranial length. The posterior fossa in modern humans was significantly smaller as a result of these changes. The posterior fossa makes up 27% of the total cerebral space in people with Chiari type I, compared to only 21% in normal persons.[3] The skulls of H. neanderthalensis were platycephalic, or flattened. Platypasia is connected to some forms of Chiari (platypasia is the flattening of the skull base).

Headaches, swallowing issues, nausea, unsteady gait, lack of hand-eye coordination, tingling and numbness in the fingers and toes, and speech issues are some of the symptoms of CM. Less frequently, people may experience symptoms like ringing or buzzing in the ears, weakness, slow or fast heartbeats, scoliosis associated with spinal cord damage, central sleep apnea, and quadriplegia. Due to the restriction of cerebrospinal fluid (CSF) outflow, this might occasionally result in non-communicating hydrocephalus.

PATHOPHYSIOLOGY

It is unknown what causes congenital CMs. The reduction or absence of development of the posterior fossa as a result of congenital or acquired diseases is the most widely acknowledged pathophysiological mechanism through which Chiari type I abnormalities take place. The abnormality could be brought on by an issue with fetal growth. It might result from exposure to teratogenic drugs while pregnant. Or it might be connected to inherited genetic issues. After birth, a person develops a type I acquired CM which is caused by excessive spinal fluid leakage from the lumbar or thoracic regions of the spine. An injury, exposure to dangerous substances, or an illness are all potential causes of this. Congenital causes include X-linked vitamin D-resistant rickets, hydrocephalus, craniosynostosis (particularly of the lambdoid suture), hyperostosis (including craniometaphyseal dysplasia, osteopetrosis, and erythroid hyperplasia), and neurofibromatosis type I. Space-occupying lesions resulting from acquired illnesses such as brain tumors or hematomas are an example.[4]

The mechanism of delayed acquired CM, which may result from traumatic brain injury, is unknown but proposed explanations include that an ectopia could exist and be asymptomatic up until a whiplash injury makes it symptomatic leading to the delayed acquired CM.[5]

DISCUSSION AND TYPES OF CMS

Suboccipital headaches and/or neck pain are the most typical presentation of Chiari I malformation. When prompted to perform the Valsalva maneuver, symptoms worsen. Ocular abnormalities, otoneurologic symptoms (dizziness, hearing loss, vertigo), gait abnormalities, and widespread fatigue are additional frequent presentations.[12] Despite being significantly less frequent, there are numerous research studies in which patients have come with solitary extremity pain or weakness. One such study outlines a patient who presented to a sports medicine clinic with unilateral shoulder discomfort and isolated muscular weakness.[11] Traditional symptoms of myelopathy include motor weakness and "dissociated sensory loss" (loss of pain and temperature sensitivity but intact fine touch and proprioception).[13]

Ataxia, dysmetria, nystagmus, and lower cranial nerve palsies (IX, X, XI, XII CN) are all cerebellar symptoms that are caused by direct compression of the medulla or cerebellum at the foramen magnum or by syringomyelia. A patient with CM may develop sleep apnea as a result of pharyngeal muscle weakness brought on by the brainstem, higher spinal cord, or lower cranial nerve compression. Patients having radiological results consistent with CM who have no clinical symptoms of the condition are a regular occurrence (incidental CM). Therefore, non-specific symptoms like widespread weariness or headaches with the characteristic pattern may not always be connected to the CMs. Most frequently in gestation or at birth, the additional variants of CMs (apart from type 0 and 1.5) are detected.

Type 0: Syringomyelia without the setting of herniation of cerebellar tonsils

Syringomyelia is a disorder in which a syrinx (a cyst or accumulation of fluid) develops in the spinal cord. Cerebrospinal fluid fills the syrinx. The cerebrospinal fluid typically surrounds and cushions the brain and the spinal cord by flowing around their periphery. But some of the extra fluid can enter the spinal cord when the flow of cerebrospinal fluid is impeded. A syrinx emerges with this. Syrinxes have a propensity to grow over time if left untreated, yet some manage to stabilize or even vanish. Injuries to the spinal cord can also result in syringomyelia. Trauma, meningitis (a spinal cord membrane infection), arachnoiditis (spinal cord membrane inflammation and scarring), hemorrhage (bleeding), and other issues may result in this damage. The spinal cord's nerves can be compressed by a syrinx. The location and compression pressure of the syrinx affected to cause the specific symptoms.

Possible signs and symptoms include urinary incontinence, numbness, tingling, temperature insensitivity, discomfort in the neck and back, scoliosis (curvature of the spine), paralysis of the hands and arms, and stiffness. Symptoms typically develop over time.

Type I: Chiari I malformation

This deformity affects fetal development and is defined by a downward displacement of the cerebellar tonsils by more than four millimeters into the cervical spinal column. Because of this displacement, the CSF that normally flows between the spinal canal and the intracranial space may get blocked. Syringomyelia/hydromyelia may go hand in hand with this type of CM. It may be a congenital condition or the result of trauma. When congenital, a child may grow up with no symptoms, however headaches and cerebellar signs are frequently present. Patients with inherited connective tissue disorders frequently have the acquired Chiari I malformation syndrome of occipitoatlantoaxial hypermobility.[6] Patients with Ehlers-Danlos syndrome or Marfan syndrome, which cause significant joint hypermobility and connective tissue weakening, are vulnerable to instability at the craniocervical junction and are therefore at risk of developing a CM. This malformation is detected more likely in adults and teens.

Thirty to fifty percent of patients with Chiari I malformation exhibit anomalies at the base of the skull and in the spine. The brainstem may get compressed as a result of the upper section of the spine is compressed into the base of the skull. The first level of the spine's (C1) bony fusion with the skull's base can also occur. Congenital fusion of the spine in the neck with potential cervical spinal cord maldevelopment is known as the Klippel-Feil deformity which can be seen in patients suffering from Chiari I malformation. Obscure cervical spina bifida (bony defect in the posterior part of the spine) may also be seen. Scoliosis also affects 16–80% of individuals with hydromyelia, especially young children whose spines are still developing. Most individuals with Chiari I malformation are typically asymptomatic. However, any of the aforementioned symptoms could manifest individually or in combination. Certain symptoms are connected to the emergence of the syrinx in Chiari I malformation: cervical pain and severe head discomfort; occipital headache that is exacerbated by coughing, sneezing, or straining; loss of pain and temperature perception in the arms and upper torso (as a result of a syrinx); loss of hand- and arm-muscle strength (as a result of a syrinx); attacks when the victim drops to the ground owing to muscle weakness (drop attacks); dizziness; spasticity; balance difficulties; distorted vision and diplopia; sleep/central apnea; and children and newborns may have modest, non-specific symptoms such as hypotonia, gross motor delay, problems eating or swallowing, failure to thrive, and opisthotonus.

Type 1.5 CM

Patients with this condition have caudal brainstem descent in addition to tonsillar ectopia. Due to recurrent syringomyelia, a sizable portion of these patients require a second surgery.[7] Patients may experience cervical tenderness, headache, dysphagia and other symptoms similar to type I CM.[8]

Type II: Arnold CM

Typically, children born with spina bifida exhibit this. The inadequate maturation of the spinal cord and/or its covering is known as spina bifida. Type II is frequently referred to as the Arnold-Chiari or "typical" CM. The brain stem and cerebellum both protrude into the foramen magnum in type II CM. In other words, the medulla, fourth ventricle, and cerebellum are all displaced downward into the cervical spinal canal. It is also characterized by the extension of the pons and the fourth ventricle. In addition to spina bifida patients, patients with myelomeningocele are also primarily affected by this type. A congenital disorder known as myelomeningocele causes the spinal cord and column to improperly close during fetal development, leaving an open spinal cord deformity at birth. Hydrocephalus, cardiovascular issues, and imperforate anus, in addition to other gastrointestinal and genitourinary issues, are some of the other abnormalities connected to myelomeningocele.[9] The traditional anatomic correlations include hydrocephalus with subsequent clival hypoplasia, tectal beaking, and low-lying torcular herophili (confluence of the sinuses). Because of the accompanying neural tube abnormality, colpocephaly may be observed.

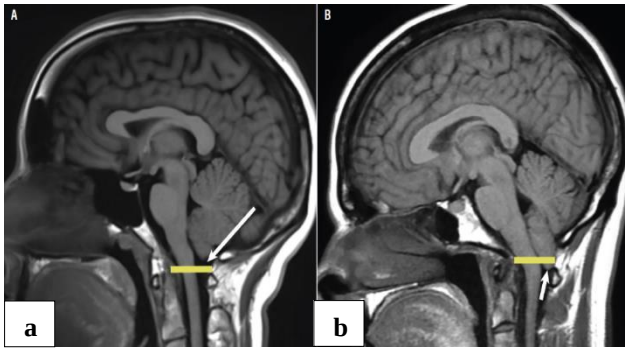

Figure 1: MRI brain of a person (a) without CM, and (b) with CM I (the cerebellar tonsils (white arrow) descend more than 5 mm below the level of the foramen magnum (yellow line)).

Myelomeningocele and hydrocephalus issues can potentially contribute to the symptoms of Chiari II malformation.[10] These signs comprise: change in breathing pattern, including apneic episodes; absent gag reflex; abrupt, sudden, and downward eye motions; and the decline in arm and leg strength.

Type III CM

This deformity comprises a type of dysraphism in which the brainstem and/or cerebellum protrude through a breach in the neck or back of the head. It is often accompanied by an occipital encephalocele, which may also contain parts of the cerebellum, brainstem, and occipital lobe that have herniated abnormally. Hydrocephalus, a tethered chord, and syringomyelia may also be present. These abnormalities are extremely uncommon and are linked to

a high likelihood of early mortality or severe neurological impairments in survivors. If treatment is started, the defect must be surgically closed as soon as possible. Shunting is also necessary to treat the frequently present hydrocephalus. There may also be more severe birth abnormalities that require considerable care. Chiari III malformation in infants can cause potentially fatal complications.[14]

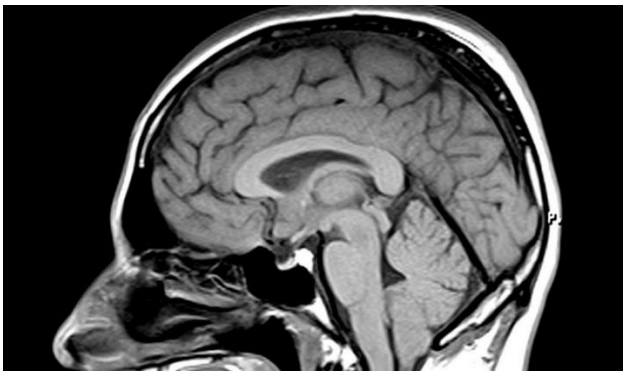

Figure 2: MRI of brain showing Arnold CM.

Type 3.5 CM

Muscatello wrote about what is thought to be the sole instance of an occipitocervical encephalocele that communicated with the stomach in 1894. This is a fascinating historical discovery, but it hasn't been seen or mentioned since. It is incompatible with life.[15]

Type IV CM

The brain stem and cerebellum are located in the posterior fossa without any connection to the foramen magnum, and the condition is characterized by a lack of cerebellar development like primary cerebellar agenesis. It is also incompatible with life.[16]

Type V CM

Type V CM is characterized by the occipital lobe ectopia into the foramen magnum and absence of cerebellar development.[17] There are just two documented occurrences of this uncommon condition in the published research, and both of them involve myelomeningocele. In light of the syndrome's postulated mechanism, this has generated debate. Some neurosurgery experts argue whether this is genuinely a distinct entity or merely a subset of the Chiari 2 malformation spectrum.[18]

EVALUATION

While Chiari II to IV are frequently first assessed by ultrasound in utero, with fetal magnetic resonance imaging (MRI) assessment being undertaken for further characterization, Chiari I are commonly evaluated by MRI in a kid or adult. The test of choice for diagnosing Chiari I is an MRI of the head and cervical spine. This will show that the tonsils of the cerebellum have descended more than five millimeters below the foramen magnum (McRae line). Additionally, a syrinx and a smaller posterior fossa may be visible.[19] It might also be necessary to add a thoracic and/or lumbar spine MRI, depending on how severe the syrinx is. CSF flow (or cine) sequences can be used in the context of ventricular dilatation to analyze CSF flow dynamics and determine whether the foramen magnum is blocked.

In the treatment of patients with Chiari I malformation, additional helpful diagnostics include: myelography which is a valuable option for patients for whom an MRI is not possible; and X-rays or computed tomography (CT) scans of the head and neck may show common concomitant bone abnormalities, particularly those at the craniocervical junction that is important for surgical planning, like basilar invagination.

Fetal sonography frequently shows the classic imaging characteristics of Chiari II and III abnormalities during the second trimester anatomical scan.

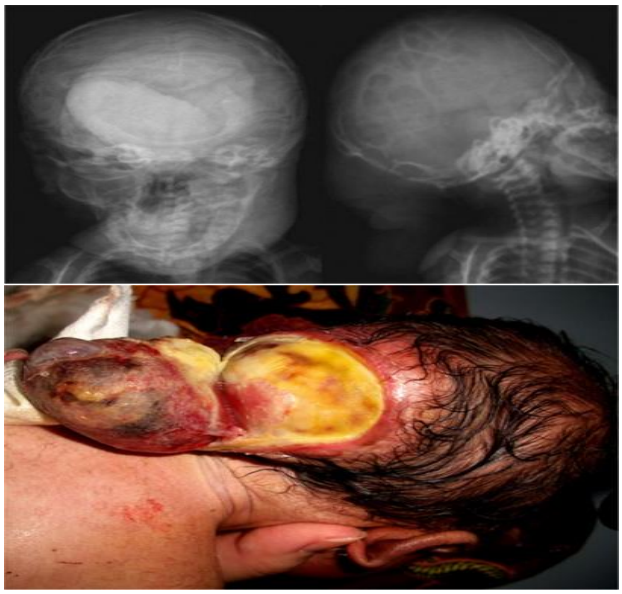

Figure 3: Type III chiari malformation: upper image shows multiple defects on calvaria (lacunar skull), lower image shows a baby affected with type III chiari malformation.

The anterior frontal calvarium's lemon sign, which is characterized by loss of the typical convex curvature and flattening or inward bowing/scalloping that gives the structure a form resembling a lemon, is one of the classic imaging findings on ultrasonography.[20] The anterior curvature of the cerebellum demonstrates aberrant morphology and there is an obliteration of the cisterna magna, making the banana sign, which is another characteristic symptom of Chiari II malformation and distal neural tube damage.[21] When assessing the posterior fossa during a prenatal exam, Chiari III will show the occipital or high cervical meningoencephalocele. Fetal

MRI can show Chiari IV and V's cerebellar hypoplasia/aplasia and help diagnose Chiari II and III's neural tube abnormalities and hindbrain herniation. Additionally, MRI provides a clearer picture of the tectal beaking present in Chiari II. Evaluation of patients with CM patients cannot be aided by laboratory investigations. However, before preparing for surgery, laboratory studies are required such as the electrocardiogram (ECG) and routine tests like complete blood count (CBC), coagulation profile, electrolyte levels, and chest X-ray.

Differential diagnosis includes: intracranial hypotension, increased intracranial pressure leading to tonsillar herniation, and normal variant cerebellar tonsillar ectopia.

TREATMENT AND PROGNOSIS

CM patients who exhibit no symptoms can be treated medically. Skeletal muscle relaxants, NSAIDs, and short usage of a cervical collar are all effective treatments for headaches and neck pain. However, research indicates that while nausea and headaches may subside with medical treatment, many symptomatic people's gaits would not. Even if they have syringomyelia, nearly 90% of Chiari type I patients may continue to be asymptomatic.

The primary method of treating CM is surgery, which aims to decompress the posterior fossa and restore CSF flow across the craniovertebral junction, reducing pressure on the cerebellum and hindbrain. Patients with proven tonsillar herniation who continue to experience symptoms may consider surgery. If there is a tonsillar herniation that is asymptomatic and either has a syrinx or not, surveillance is advised until symptoms appear. When surgery is done within two years of symptom onset, the outcomes are better.[22]

A syrinx is known to cause a variety of distinct indications and symptoms, such as dysesthesia, algothermal dissociation, spasticity, and paresis. These are significant indicators that patients with CM type II require decompressive surgery.[23] Patients with type II diabetes have severely damaged brain stems and rapidly deteriorating neurological function. In order to relieve pressure, decompressive surgery entails removing the lamina of the first cervical vertebra, and occasionally the second or third, as well as a portion of the skull's occipital bone. An additional spinal fluid flow may be achieved with a shunt. A dural graft is typically used to fill the increased posterior fossa because this operation typically entails opening the dura mater and expanding the area beneath. A minority number of neurosurgeons hold the alternative theory that untethering the spinal cord lessens the pressure that the brain experiences against the foramen magnum, preventing the need for decompression surgery and related damage.

With only reports on a small number of individuals, this method is, however, substantially less well-documented in the medical literature. Alternative spinal surgery carries some danger as well.

Decompression surgery may result in complications. These include hemorrhage, meningitis, CSF fistulas, occipito-cervical instability, and pseudomeningeocele in addition to harm to the brain and spinal cord structures. Hydrocephalus and odontoid retroflection-induced brain stem compression are uncommon post-operative problems. A cerebellar "slump" may also result from a prolonged CVD brought on by a wide opening and large duroplasty. Cranioplasty can cure this complication.[24]

It is possible for the brainstem to experience irreversible compression from the front (anteriorly or ventrally), which can lead to a smaller posterior fossa and accompanying CM. An anterior decompression is necessary in these circumstances. The most common method is to remove the bone, usually the odontoid, that is crushing the brainstem through the mouth (transoral). The brainstem decompresses as a result, making more space for the cerebellum and decompressing the CM.[25]

The prognosis for Chiari I is good, but it also relies on whether there are any neurological abnormalities that exist. The majority of people with no neurological impairments fare very well.[26] The neonatal in-hospital mortality rate of Chiari II is 3%, while the 3-year death rate is 15%. Those who survive may develop progressively worsening motor impairment. It is advised to continue monitoring for shunt placement issues or failure. The prognosis is poor and frequently grim in the more severe Chiari forms, with early mortality. Chronically weak or gait-impaired people typically don't get better, and their prospects are uncertain.[27]

COMPLICATIONS OF CM

The complications of CM include: wound infection, CSF leak, meningitis, pseudomeningocele, epidural Haemorrhage, vertebral artery damage, hydrocephalus, syringomyelia, and apnea.

CONCLUSION

The CM is a disorder where the spinal canal is invaded by neural tissue. It happens when the brain is pressed against and forced downward by an abnormally shaped or tiny portion of the skull. CM is rare, although more imaging tests are being used, which has increased the frequency of diagnoses. As the skull and brain mature, CM type 1 occurs. Signs and symptoms may not show up until late adolescence or adulthood as a result. Types 2 and 3 of the pediatric CM are present from birth (congenital). The form, degree, and accompanying symptoms of a CM all affect how it is treated. Treatment options include routine observation, prescription drugs, and surgery. The importance and impact of CM is now comprehended more as a result of the growing use of MRI for testing children and people with mild neurological illness. The

dangers of any surgery are undoubtedly reduced by doing this, and those who have this illness will experience less long-term morbidity as a result.

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