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Arnold Chiari Malformation

Authors

Joaquin A. Hidalgo[1]; Matthew Varacallo[2].

Affilations

[1] University of Mississippi
[2] Department of Orthopaedic Surgery, University of Kentucky School of Medicine

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Introduction

Arnold-Chiari, also know as Chiari malformation, is the name given to a group of deformities of the hindbrain (cerebellum, pons and medulla oblongata). Issues range from herniation of the posterior fossa contents outside of the cranial cavity to absence of the cerebellum with or without other associated intracranial or extracranial defects such as hydrocephalus, syrinx or spinal dysraphism. [1],[2],[3]

Classification

It is based on the morphology of the malformations as evidenced radiologically or at autopsy.

- *Chiari I*: Greater than 5-mm descent of the caudal tip of cerebellar tonsils past the foramen magnum
- *Chiari II*: Brainstem, fourth ventricle, and greater than 5-mm descent of the caudal tip of cerebellar tonsils past the foramen magnum with spina bifida.
- *Chiari III*: Herniation of the cerebellum with or without the brainstem through a posterior encephalocele.
- *Chiari IV*: Cerebellar hypoplasia or aplasia with normal posterior fossa and no hindbrain herniation.

Etiology

There are multiple proposed theories including molecular, hydrodynamic, and mechanical.[4]

It is likely that different mechanisms can have the same resulting Chiari malformation.

Chiari Type I Malformation

Genetic syndromes related or more commonly idiopathic reduced volume of the posterior fossa leads to a displacement of the cerebellar tonsils into the spinal canal.

Chiari Type II Malformation

Invariably results in the setting of myelomeningocele. Leakage of cerebrospinal fluid (CSF) to the myelomeningocele yields herniation of the hindbrain into the spinal canal, which disturbs the cerebrospinal fluid normal flow and subarachnoid adhesions.

Epidemiology

Chiari malformation is type 1 is the most common type and occurs in approximately 1 in 1000 births.[5]

There is a slight female predominance of cases (1.3 to 1).

Type 2 malformation is associated with neural tube defects, especially myelomeningocele, in almost all cases.

Pathophysiology

Neurologic signs and symptoms can arise from 2 mechanisms:

- Direct compression of neurological structures against the surrounding foramen magnum and spinal canal
- Syringomyelia or syringobulbia development
    - The obstruction of cerebrospinal fluid (CSF) outflow eventually results in syrinx development
    - Fluid-filled cavities (syrinx) develop within the spinal cord or brainstem, resulting in neurologic symptoms as the cavity expands.[6]

In patients with the Chiari type I malformation, the bones of the skull base often are underdeveloped, which results in a reduced volume of the posterior fossa, the volume of which is inadequate to contain the entire cerebellum, thus cerebellar tonsils are displaced into the cervical canal.

The posterior fossa in Chiari type II malformation is even smaller than in Chiari I malformation. The cerebrospinal fluid (CSF) cisterns are poorly developed due to lack of brain cistern expansion as a consequence of in-utero derivation CSF circulation to the neural tube defect, all of which results in hindbrain structures downward herniation with subsequent compression of these structures against the foramen magnum. [6]

In both type I and II, there is CSF-flow obstruction by the foramen magnum crowding, and consequently, hydrocephalus and/or syringomyelia formation are possible over time.

History and Physical

In Chiari type I malformation the most common presentation is suboccipital headaches and or neck pain (80%). Symptoms are exacerbated when asked to perform the Valsalva maneuver. Other common presentations include ocular disturbances, otoneurologic symptoms (dizziness, hearing loss, vertigo), gait ataxia, and generalized fatigue. Although much less common, the literature reports multiple case studies in which patients have presented with isolated extremity pain or weakness, one such report including a presentation of unilateral shoulder pain with isolated muscle weakness presenting to a sports medicine clinic [7].

Myelopathy classically presents with “dissociated sensory loss” (loss of pain and temperature sensation, preserved fine touch and proprioception) and motor weakness. [8],[9]

Cerebellar signs, including ataxia, dysmetria, and Nystagmus, lower cranial nerve deficits (IX, X, XI, XII CN) either from direct compression of the cerebellum or medulla at the foramen magnum or from syringomyelia or syringobulbia.

Sleep apnea can occur in a patient with Chiari malformation due to a weakness of pharyngeal muscles elicited by the brainstem, upper spinal cord, or lower cranial nerve compression.

It is not an uncommon scenario to find patients with radiological findings compatible with Chiari malformation with no clinical manifestations of the disease (incidental Chiari malformation). Therefore, unspecific symptoms such as generalized fatigue or classic pattern migraines are not necessarily related to the Chiari malformation.

Evaluation

MRI of the Head and Cervical Spine

It is the test of choice. Demonstrating the tip of the cerebellar tonsils 5 mm below the foramen magnum, associated possible syrinx, and regional malformations.[10]

Other useful tests in the management of patients with Chiari malformation include:

- *Myelography*: Of special value as an alternative in patients in who an MRI cannot be obtained

- *CT or x-rays of the neck and head:* May reveal common associated bony defects particularly of the craniocervical junction relevant for surgical planning
- *Cine-MRI:* Useful in the evaluation of cerebrospinal fluid dynamics. May demonstrate blockage of flow at the foramen magnum. However, it is not widely utilized due to the modality's uncertain accuracy.

Laboratory studies are not of help in the evaluation of patients with Chiari malformation. However, laboratory studies are needed during planning for surgery. Routine studies like the complete blood count (CBC), coagulation profile, electrolyte levels, chest X-ray, and ECG will suffice.

Treatment / Management

The main treatment for Chiari malformation is surgical with the goal of re-establishing the CSF flow across the craniovertebral junction and decompressing the nervous system elements.[11],[4]

Medical Management

Patients with Chiari malformation and who have no symptoms can be managed medically. Headaches and low neck pain can be treated with muscle relaxants, NSAIDs, and temporary use of a cervical collar. However, studies show that while a headache and nausea may improve, in many symptomatic patients there will be no improvement in gait with medical management. Close to 90% of patients with Chiari type I may remain asymptomatic even if they have syringomyelia.

Indications

Surgery is recommended for patients complaining of the classic pattern of symptoms and confirmed tonsillar herniation.

In the setting of tonsillar herniation without clinical manifestations, observation is recommended, surgery is recommended if symptoms develop.

The general consensus regarding incidental findings of a syrinx in the setting of a normal comprehensive examination includes observation alone.

Better surgical results are seen when surgery is performed within 2 years of symptoms onset.

Surgical Techniques

Posterior fossa decompression: suboccipital craniotomy with or without the following:

- C1 and C2 laminectomy with or without[12]
- Dural opening/patching with or without
- Tonsilar cauterization

In general, patients who are asymptomatic without syringomyelia and who had an incidental diagnosis following an MRI study should not undergo surgery. This group can be managed with close monitoring and medical treatment.

Contraindications to Surgery

When other pathologies cause the tonsillar herniation, for example, a malignant or benign lesion or CSF hypotension syndrome, besides Chiari malformation, decompression of the foramen magnum is contraindicated.

Prognosis

For Chiari type I malformation the prognosis is good, but it also depends on the presence of preexisting neurological deficits. Most patients who have no neurological deficits have an excellent outcome.[13],[5]

Individuals who have chronic weakness or gait problems usually do not improve and their prognosis is guarded.[14]

Complications

- Pseudomeningocele
- CSF leak
- Meningitis
- Wound infection
- Lower brainstem malfunction
- Epidural hematoma
- Apnea
- Vertebral artery injury

Postoperative and Rehabilitation Care

In the postoperative period, monitoring for CSF leak is vital. Some patients may develop a pseudomeningocele, which may require drainage.

Exercise and heavy lifting should not be done for at least 3 to 4 weeks after the procedure.

Most patients require 6 to 8 weeks to recover from surgery and reverse any major neurological deficit fully.

Repeat MRI is necessary to ensure that the syrinx has responded to the treatment.

Pearls and Other Issues

Arnold-Chiari, also known as Chiari malformation, is the name given to a group of deformities of the hindbrain (cerebellum, pons and medulla oblongata).

Issues range from herniation of the posterior fossa contents outside of the cranial cavity to absence of the cerebellum with or without other associated intracranial or extracranial defects such as hydrocephalus, syrinx, or spinal dysraphism.

For Chiari type I malformation, the prognosis is good, but it also depends on the presence of preexisting neurological deficits. Most patients who have no neurological deficits have an excellent outcome.

A controversial subject in clinical management entails the diagnosis in the setting of possible sport-specific participation.

Individuals who have chronic weakness or gait problems usually do not improve, and their prognosis is guarded.

Enhancing Healthcare Team Outcomes

Arnold Chiari malformations are relatively common and represent a spectrum of hindbrain anomalies. Depending on the severity of the malformation, the individual may be asymptomatic or have severe neurological symptoms. While the patients are often managed with decompressive surgery, the nurses are responsible for looking after these individuals. Hence, the nurse must be aware of the potential post-surgical complications and their presentation. The prognosis for most patients with a Chiari malformation is good, but it also depends on the initial neurological presentation. Those patients with mild neurological deficits tend to have good outcomes but those with moderate to severe symptoms, tend to have a guarded prognosis. The surgery is also associated with a number of complications of which the most common is a pseudomeningocele. A few individuals may have a persistent syrinx and may require a shunt. [15](Level III)

Questions

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