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Chiari Malformations: Historical Background, Anatomical Forms and Treatment Approaches

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Review

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ABSTRACT

Chiari malformations (CM) refer to a series of anomalies characterized by the descent of cerebellar tonsils into the cervical spinal canal. These malformations can be associated with abnormalities such as syringomyelia, hydrocephalus, spina bifida, and scoliosis. Additionally, cranio-cervical junction anomalies, endocrinopathies, craniosynostosis, and syndromic disorders are also linked to CM. The treatment of CM is surgical, and there is no known medical therapy. Patients diagnosed with CM are typically advised to undergo surgical treatment or follow-up. Although surgical intervention is supported in the literature, debates exist regarding which procedure is most suitable and when surgery should be performed. In this article, we will examine the historical background of CM, its anatomical forms, pathophysiology, clinical presentation, relationship with other diseases, and diagnostic procedures in the light of the literature.

Keywords: Arnold Chiari Malformation, history, etiology, diagnosis, treatment

Chiari Malformasyonları: Tarihsel Arka Plan, Anatomik Formlar ve Tedavi Yaklaşımları

Derleme

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ÖZET

Chiari malformasyonları (CM), serebellar tonsillerin servikal spinal kanala inmesiyle karakterize edilen bir dizi anomaliyi ifade eder. Bu malformasyonlar, siringomiyeli, hidrosefali, spina bifida ve skolyoz gibi anormalliklerle ilişkilendirilebilir. Ayrıca, kraniyoservikal bileşke anomalileri, endokrinopatiler, kranyosinostoz ve sendromik bozukluklar da CM ile bağlantılıdır. CM'nin tedavisi cerrahidir ve bilinen bir tıbbi tedavisi yoktur. CM tanısı alan hastalara genellikle cerrahi tedavi veya takip önerilir. Cerrahi müdahalenin literatürde desteklendiği görülse de hangi prosedürün en uygun olduğu ve ameliyatın ne zaman yapılması gerektiği konusunda tartışmalar bulunmaktadır. Bu makalede, CM'nin tarihsel arka planı, anatomik formları, patofizyolojisi, klinik sunumu, diğer hastalıklarla ilişkisi ve literatür ışığında tanı prosedürleri incelenecektir.

Anahtar Kelimeler: Arnold Chiari Malformasyonu, Öykü, Etiyoloji, Tanı, Tedavi

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Introduction

Chiari Malformation is a group of conditions characterized by the descending part of the posterior brain structures through the foramen magnum into the cervical spinal cord.[1] This condition was first described by J. Cleland in 1883. In 1894, German pathologist Julius A. Arnold identified and published a case of posterior brain herniation in a myelodysplastic patient in Heidelberg. Subsequently, Hans von Chiari classified these herniations into grades, creating a classification system. Chiari initially described three types of malformations and later added a fourth type, which does not involve herniation of the posterior fossa structures. This classification was completed in 1896 by Hans Chiari, leading to the well-known classic four types of Chiari Malformation.[2] Over the years, different variations of Chiari Malformation have been identified. These include Chiari Malformation Type 0 (Chiari-like malformation), Type 1.5, Type V, and Complex Chiari.[3]

A significant milestone in the surgical treatment of Chiari Malformation was set in 1932 by Cornelis Joachimus van Houweninge Graftdijk, a Dutch professor of neurosurgery. Graftdijk performed the first Chiari operation, removing the suboccipital bone and excising the cerebellar tonsils. Some losses were also reported in these early surgical interventions.[4]

In 1941, studies conducted by List,[5] and in the same year by Adams, Schatzki, and Scoville[6] first demonstrated obstruction at the level of the foramen magnum using myelography, marking a significant advancement in the diagnosis of Chiari disease. With the widespread adoption of Computed Tomography (CT) and Magnetic Resonance Imaging (MRI) technologies, the diagnosis of Chiari Malformation has become even more accessible.

Physiopathogenesis

Several theories have been proposed to understand the etiology of Chiari Malformation (CM), but none have been sufficient to fully explain this complex condition on their own. The origin of CM, whether as a continuation of an embryological anomaly or as a result of a pathology developed later, remains unclear. The following are the main theories in this regard:

Traction Theory: According to this theory, conditions located caudally, such as meningomyelocele, cause the spinal cord to adhere to surrounding tissues and pull the hindbrain structures downward. This process impedes the upward migration of the tonsils during the 9th week of intrauterine life.[7] However, this theory does not explain the upward movement of the cerebellum towards the tentorium.[8]

Developmental Arrest Theory: This theory posits that dysgenetic development in the brainstem is the primary pathology. A developmental arrest during embryogenesis prevents the formation of the pontine flexure, leading to the elongation of the brainstem and the descent of posterior fossa structures into the spinal canal.[9]

Hydrodynamic Theory: Proposed by Gardner in 1950, this theory suggests that pulsatile forces in the supratentorial ventricular region's choroid plexus and the area of the 4th ventricle differentially affect brain development. When supratentorial forces are dominant, the tentorium is pushed caudally, causing compression of posterior fossa structures.10 If the pulsatile forces of the 4th ventricle dominate, it results in the development of Dandy-Walker anomaly.[11]

Overgrowth Theory: In 1932, van Houweninge Graftdijk proposed that the vertebral column grows faster than the spinal cord, leading to the migration of infratentorial structures towards the occipital foramen.[12]

Small Posterior Fossa Theory: This theory explains that a primary paraxial mesodermal insufficiency results in the bony structures of the posterior fossa being smaller than normal, leading to the herniation of the brainstem and cerebellum into the cervical canal.[13]

Neurodystrophic (Distrophic) Theory: Described by Osaka and colleagues in 1978, this theory suggests that developmental disorders of the lumbar spinal cord, similar to conditions like meningomyelocele, lead to cerebrospinal fluid (CSF) leakage. This leakage causes a decrease in CSF pressure, which in turn allows the cerebellar tonsils to sag into the upper cervical canal. However, this theory does not explain Type 1 Chiari Malformation (CM1).[13,14]

These theories, while contributing significantly to the understanding of the complex nature and etiology of Chiari Malformation, each explain only specific aspects of the disease. A comprehensive understanding requires integrating these theories.

Diagnosis

There are several imaging and testing methods used for the diagnosis of Chiari Malformation (CM), playing a critical role in its accurate identification and treatment.

Cervical Magnetic Resonance Imaging (MRI): MRI is the most important imaging method for diagnosing Chiari Malformation.[15] Typically, a descent of the cerebellar tonsils more than 5 mm below the foramen magnum is considered pathological. However, some sources also consider a herniation of 0 or 2 mm as pathological.[16]

Computed Tomography (CT) and Direct Radiographs: These imaging methods are preferred in patients suspected of having cranio-cervical bone anomalies. If MRI is contraindicated, metrizamide-enhanced CT imaging can be used as an alternative option.[8,17]

Cine-Mr Imaging: This is used to assess cerebrospinal fluid (CSF) circulation in CM1 patients with syringomyelia. Cine-MR imaging can reveal that while CSF flow in the anterior region is normal in Chiari patients, there is reduced or absent flow in the posterior region. Four-dimensional MRI flow techniques have shown the potential to reveal complex flow characteristics through qualitative and quantitative flow analysis.[15,18,19]

Ultrasonography (Usg): Used in infants for the diagnosis of Chiari and the detection of syringomyelia.

Routine prenatal ultrasonographic examinations can identify developmental anomalies such as Chiari malformation and spinal dysraphism early on.[18,19]

Neurophysiological Tests: These tests can assess compressions of the posterior fossa and spinal cord. However, it has been reported that neurophysiological tests are not always functional in children.[20]

These imaging and testing methods are vital for the accurate diagnosis and appropriate treatment planning of Chiari Malformation. Each method illuminates different aspects of the disease, aiding in the best possible treatment of patients.

Chiari Malformation Types and Clinical Findings

Chiari Malformation is divided into various types, and each type has its own unique features and treatment needs.

Chiari Malformation Type 1 Features and Clinical Findings

Definition and Frequency: CM1 is characterized by the herniation of the cerebellar tonsils into the cervical canal by 5 mm or more. It usually occurs sporadically, and genetic transmission is rare.[21] It is more common in women and most often manifests between the ages of 30 and 50.[21,22] Cranial and cervical pathologies may also accompany CM1. CM1 is found in about 0.9% of the adult population and 0.6% of the pediatric population, with one-third of patients exhibiting clinical symptoms.[23]

Clinical Symptoms: CM1 patients may present with symptoms ranging from simple head and neck pain and numbness in the extremities to more serious complaints such as difficulty swallowing and sudden respiratory arrest.[24] The level of cerebellar tonsil herniation can influence the severity and variety of clinical findings, though this is not always the case.[25] Clinical symptoms may occur due to compression of the brain stem, spinal cord, lower cranial nerves, and cerebellum caused by volume loss in the posterior fossa and the descent of the cerebellar tonsils.[26,27]

Classification of Findings: Milhorat and colleagues have classified the clinical findings of CM1 into five main groups: Suboccipital headache, ocular disorders, neuro-otological disorders, brain stem-related clinical findings, and spinal cord findings.[28]

Pain: Headache is the most common complaint in 80-90% of CM1 patients.[22] Pain, particularly concentrated in the occipital and upper cervical region and triggered by Valsalva maneuvers, is typical.[22]

Cerebellar Findings: Approximately 30-52% of CM1 patients exhibit cerebellar signs. These include balance disturbances, gait, and coordination problems, dysmetria, dysdiadochokinesia, dysarthria, ataxia, and nystagmus.[29,30]

Lower Cranial Nerve and Brainstem Involvements: These symptoms are less common and may include voice thickening, dysphagia, facial sensory deficits, and sleep apnea.[29-31]

Spinal Cord Findings: The common occurrence of clinical findings related to the spinal cord (63%) can be explained by the frequent radiological association of CM1 with syringomyelia (30-70%). Motor and sensory loss are common.[26,27]

Ophthalmological and Otological Findings: Symptoms such as blurred vision, nystagmus, diplopia, and tinnitus have also been reported among CM1 patients.[30]

Cognitive Complaints and Mood Disorders: CM1 patients may experience psychiatric disorders such as distractibility, depression, and general lethargy. Although the pathogenesis of these complaints, which may benefit from surgery, is not fully explained, they have been suggested to possibly be a form of "cerebellar seizure".[32]

Systemic Problems: Systemic symptoms like chest pain, shortness of breath, hiccups, postural hypotension, and syncopal attacks have also been reported.[26,27,31]

Other Types of Chiari Malformation

Chiari Type II: Commonly seen in children and often associated with spinal dysraphism. It is characterized by the descent of the brainstem and posterior fossa structures into the cervical canal. Also known as Arnold-Chiari syndrome.[21,33]

Chiari Type III: The rarest type, characterized by the herniation of the cerebellum and brainstem into an encephalocele sac. It has the worst prognosis.[21]

Chiari Type IV: Defined by severe cerebellar aplasia or hypoplasia. The posterior fossa volume is reduced, but there is no descent into the spinal canal. Despite poor radiological images, patients exhibit mild to moderate neurological deficits. It does not cause significant symptoms and does not require treatment.[9]

Chiari Type V: Characterized by cerebellar agenesis and herniation of the occipital lobes into the cervical canal.[34]

Chiari Type 0: Characterized by the presence of syringomyelia without tonsillar herniation and improvement of clinical symptoms following posterior fossa decompression.[16]

Chiari Type 1.5: Defined as a type of Chiari where, in addition to the herniation of cerebellar tonsils, as seen in Chiari Type 2, the brainstem, vermis, and IV ventricle are also involved, but without accompanying spinal dysraphism. Similar to CM1, it is observed in the adult age group.[33]

The term "Complex Chiari," as defined by Brockmeyer in 2011, refers to a more complex condition seen in patients with Chiari Malformation Type 1 (CM 1). This condition encompasses cases of CM 1 accompanied by one or more radiographic findings such as brainstem herniation, retroflexed odontoid, basilar invagination, abnormal clivus-cervical angle, occipitalization of the atlas, syringomyelia, or scoliosis. Brockmeyer noted that Posterior Fossa Decompression (PFD) alone may not be sufficient in these complex CM 1 cases. Additional treatments such as odontoid resection and occipitocervical fusion may be necessary.[3] This approach has been supported in subsequent studies.[35]

Treatment methods vary based on the patient's symptoms and the type of malformation. Surgical intervention is preferred, especially in CM1 cases with significant symptoms and accompanying syringomyelia.

Pathologies Accompanying CM1

Syringomyelia is a condition characterized by the formation of longitudinal cystic cavities in the spinal cord, leading to compression and various neurological issues. Though more common in adults, it can also occur in children and is often associated with anomalies of the spine and cervicomedullary junction. Known by various names such as syrinx, hydromyelia, syringohydromyelia, and intramedullary cyst, syringomyelia most frequently appears in the lower cervical region but can occur anywhere along the spinal cord.[37] This condition is predominantly congenital and shows more clinical manifestations between the ages of 25-40, though it is rarer in children, with distal syringomyelia being more common in this age group. Slightly more prevalent in males, syringomyelia is a common pathology accompanying CM1, found in 50-80% of CM1 patients, typically beginning in the cervical region.[22,37] Syringomyelia is a slowly progressing disease with variable symptoms.[36] In adults, syringomyelia can present various clinical symptoms, while in children, spinal anomalies and scoliosis are significant findings.[37,38]

Scoliosis is a common condition in patients with CM1 and syringomyelia. About 15-30% of CM1 patients have scoliosis.[22,30]

Hydrocephalus is frequently seen in CM1 patients, with an incidence of approximately 7-10% at diagnosis.[37,39] Hydrocephalus is a significant complication in CM1 patients and can be treated with shunt surgeries or endoscopic third ventriculostomy (ETV).[37,38]

Posterior Fossa Arachnoid Cysts have been associated with CM1 and syringomyelia, and surgical intervention may be required in these cases. These surgeries aim to increase cerebrospinal fluid (CSF) flow and relieve congestion at the level of the foramen magnum.[40]

Craniosynostosis was first reported in CM1 cases by Saldino and colleagues.[41] It occurs due to the premature closure of skull bones and has been observed that surgeries for craniosynostosis in CM1 patients can correct tonsillar herniation and clinical symptoms.[41]

Hyperostosis, or excessive thickening of bones, particularly when occurring in the posterior fossa region, can lead to a reduction in the volume of this area and contribute to the herniation of the cerebellar tonsils. There is a possibility of CM1 occurring in Paget's disease.[42]

Osteopetrosis, typically seen in late childhood or adulthood, is characterized by thickening of the skull and neurological symptoms due to the narrowing of cranial foramina. It has been suggested that excessive thickening of the posterior fossa bones in osteopetrosis can lead to CM1.[43]

Bone Mineral Deficiency, especially in familial vitamin D-resistant rickets, indicates that CM1 can commonly occur in association with thickening of the posterior fossa bones. Low serum phosphate levels can trigger bone growth and thickening, leading to the narrowing of the foramen magnum and the development of CM1.[44]

Endocrine Disorders can be associated with CM1. In cases of growth hormone deficiency, CM1 has been reported in 5-20% of patients. Acromegaly, causing excessive bone growth and thickening of the posterior fossa bones, can contribute to the development of CM1.[45] Similarly, achondroplasia can lead to a reduction in the volume of the posterior fossa and the development of CM1.[42,45]

Basilar Invagination, one of the most common craniocervical bone anomalies, is defined by the odontoid process extending more than 5 mm above the Chamberlain line.[46] A study reported a 14.2% association with CM1.[46] In cases of CM1 combined with basilar invagination, performing only foramen magnum decompression (FMD) may lead to instability or lack of improvement in patients.[47] In such cases, if craniovertebral instability is present, stabilization in conjunction with decompression is recommended.[47] Another approach can be anterior dens resection followed by FMD and posterior stabilization.[48]

A study reported significant improvement in patients with CM1 and basilar invagination after FMD and stabilization.[47] For patients with basilar invagination without anterior compression, FMD alone may suffice.[46]

Atlantoaxial Dislocation (AAD) can occur in conjunction with CM1. AAD should be considered in CM1 patients who exhibit symptoms such as neck pain that worsens with movement, advanced motor deficit, spasticity, posterior column signs, and sphincter disturbances.[49]

Atlas Occipitalization is frequently encountered in CM1, especially in cases with basilar invagination.[49] This condition involves an abnormal fusion between the atlas and the occipital bone. It has been reported that brainstem compression symptoms are more common in patients with CM1 and atlas occipitalization than in typical CM1 patients.[50]

Klippel-Feil Syndrome is a condition characterized by the abnormal fusion of spinal bones and is associated with CM1 in about 3.3-5% of cases.[48] Due to the frequent co-occurrence of Klippel-Feil Syndrome and CM1, it is thought that these conditions might share a common genetic disorder.[51] Klippel-Feil Syndrome primarily occurs in the upper cervical region and often presents with Atlantoaxial Dislocation (AAD).[49]

Neurofibromatosis Type 1 is an autosomal dominant neurocutaneous syndrome characterized by various skin lesions, neurological findings, and tumor formation. It is one of the most commonly associated neurocutaneous syndromes with CM1. CM1 can be found in approximately 5% of patients with neurofibromatosis type 1.[52] It is believed that this syndrome contributes to the development of CM1 by causing a halt in the development of the posterior fossa.[51] A connection between the SUZ12 gene, which codes for the SUZ12 protein, and NF1 and CM1 has been reported, and this gene is located on chromosome 17.[51]

Rare diseases that are associated with Chiari syndrome include hereditary connective tissue disorders, bone dysplasias, transverse sinus stenosis, spina bifida, intracranial hypotension, Klippel-Trenaunay syndrome,

Morning Glory disk anomaly, dehiscence of the semicircular canals, Rubinstein-Taybi syndrome, and Gorham's disease of the skull base.

These associations underscore the complexity of Chiari syndrome and the need for comprehensive evaluations in patients, as they may present with a spectrum of related conditions that impact diagnosis and treatment strategies.

Treatment

Treatment and follow-up of CM1 remain subjects of various debates. Primarily, if the patient has other conditions like hydrocephalus or Craniospinal Junction Abnormalities Classification (CSJAC), it is recommended to address these emergencies or conditions first before focusing on correcting CM1. Approximately 7-10% of CM1 patients are found to have hydrocephalus.[39] In a study by Klekamp, only 9.5% of 644 CM1 patients were followed up without surgery, indicating that most CM1 patients require surgical intervention.[37]

The natural course of Syringomyelia (SM) associated with CM1 is not fully understood. Nishizawa and colleagues followed 9 patients without significant clinical symptoms and neurological findings for 11 years, reporting that only one required surgery due to clinical deterioration.[38] In untreated cases of SM, irreversible cervical myelopathy can develop in the cord.[53]

The consensus is to recommend surgical intervention for all CM1 cases accompanied by a syrinx. However, studies have observed that in asymptomatic cases with a syrinx, many patients did not show growth in their syrinx cavities when followed without surgery.[53,54] Therefore, asymptomatic CM1 patients, even with a syrinx cavity, can be managed without surgery. However, surgical treatment is commonly preferred in patients with progressive scoliosis or clinical symptoms.[55]

In patients without a syrinx cavity, if there is minimal tonsillar herniation and non-life-limiting headaches, monitoring is advised. However, surgical treatment is recommended in cases of life-limiting headaches, respiratory distress, or cranial nerve involvement.[30] This underscores the need for individual assessment of each CM1 patient based on their unique condition and symptoms.

The standard surgical treatment for Chiari Malformation Type 1 is known as Foramen Magnum Decompression (FMD). This procedure aims to relieve pressure on the brainstem and normalize the flow of cerebrospinal fluid (CSF). FMD involves removing part of the margin of the foramen magnum and often includes the removal of the C1 lamina. If the tonsillar herniation extends down to the C2 level, the upper part of the C2 lamina may also need to be removed.[32]

Foramen Magnum Decompression (FMD) can be performed using two main methods:

Foramen Magnum Decompression and C1 Laminectomy with Vertical Dural Incisions: This method usually preserves the integrity of the dura mater. Vertical incisions on the dura are made to relieve the pressure without opening it.[22,56]

FMD and C1 Laminectomy with Dural Opening and Duraplasty: This method provides a more extensive decompression by opening the dura mater and enlarging it with Duraplasty. It is used to expand the dura mater for more comprehensive decompression.[22,56]

Recently, new treatment approaches have been suggested for CM and syringomyelia, especially when associated with atlantoaxial instability. Notably, Goel has emphasized the necessity of C1-C2 stabilization in the treatment of CM and syringomyelia.[57] This approach could be a significant alternative, particularly for patients with instability in the atlantoaxial joint. Such innovative treatment methods offer new perspectives in the treatment of CM1 and syringomyelia, playing a crucial role in providing more specific interventions tailored to the patient's conditions.[57-58]

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