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CHIARI MALFORMATION TYPE 1 AND META ANALYSIS OF PERSISTENT/RECURRENT SYRINGOMYELIA AFTER DECOMPRESSION

Introduction

Chiari malformations are a heterogeneous group of disorders (generally congenital) that are defined by anatomic anomalies of the cerebellum, brainstem, and craniocervical junction, with downward displacement of the cerebellar structures. These malformations are often associated with spinal cord cavitations (i.e., syringomyelia). In most cases, the posterior fossa is small, and neural elements are crowded and impacted at the foramen magnum [1].

Goal

In this article, a comprehensive review of Chiari Malformation Type 1 is provided including its clinical presentation, diagnosis, and management options. A comparative meta-analysis of patients with persistant syringomyelia symptoms after foramen magnum decompression surgery are discussed

Material and methods of research

Available search engines were utilized to identify publications dealing with recurrent or residual syrinx after Chiari decompression and/or management of the syrinx. Rates of residual or recurrent syrinx were extracted, and management strategies were recorded. Overall strength of evidence was quantified

Results of the research and their discussion

Description

CM-I is characterized by cerebellar tonsils that are abnormally shaped and downwardly displaced below the level of the foramen magnum. The normal cerebellar tonsils may lie up to 3 mm below the foramen magnum in adults. In general, tonsils lying 5 mm or more, below the foramen magnum on neuroimaging are consistent with a Chiari malformation.

Syringomyelia (syrinx) – Syringomyelia, often accompanied by scoliosis, occurs in approximately 35 percent of patients with symptomatic CM-I.

Among patients with neurologic deficits due to a syrinx, the earliest sign is loss of the superficial abdominal reflexes. Other signs and symptoms include gait disturbance, radicular pain, dysesthesia, paroxysmal pruritus, upper motor neuron signs in the legs, and lower motor neuron signs maximally in the arms in those with a cervical syrinx, the most common location associated with CM-I.

Symptoms of Chiari I malformations

Many people with a Chiari I malformation will not have any symptoms. Sometimes they're only found after an MRI scan of the brain is carried out for another reason.

If symptoms do develop, they can include headaches – these are usually felt at the back of the head and may be brought on or made worse by coughing, straining, sneezing or bending over, Neck pain, dizziness and balance problems, muscle weakness, numbness or tingling in the arms or legs, blurred vision or double vision and sensitivity to light, difficulty in swallowing, tinnitus, difficulty sleeping (insomnia) and clinical depression.

If there is case associated with syringomyelia, there can be problems with using their upper limbs, difficulty walking, pain, and problems with bladder or bowel control.

Diagnosis

The diagnosis of Chiari malformations is based upon neuroanatomy. There are no biomarkers in blood, cerebrospinal fluid (CSF), or cultured tissue to confirm the diagnosis. Thus, neuroimaging is of prime importance.

Imaging

MRI of brain and spinal cord – Magnetic resonance imaging (MRI) of the brain and the whole spinal cord is the best imaging modality for evaluation of Chiari malformations. Sagittal, coronal, and axial views of the brain along with sagittal and axial images of the entire spinal cord (cervical, thoracic, and lumbar) using T1- and T2-weighted MRI sequences are useful for detecting cerebellar and brainstem displacement, associated craniocervical junction abnormalities, and hydro syringomyelia.

CSF flow imaging – We suggest obtaining a phase contrast cine MRI for patients with CM-I to look for impairment of CSF flow across the foramen magnum. This information can be used to select patients for surgical decompression of the foramen magnum in order to establish normal CSF flow.

CT – For patients who cannot have MRI, high-resolution computed tomography (CT) scan with sagittal reconstructions can be used to make the diagnosis of Chiari malformation. CT, especially thin section multiplanar CT with reformatted images, retains importance in the evaluation of the associated bony abnormalities.

Fetal ultrasound – In some cases of fetal ventriculomegaly, a Chiari malformation can be diagnosed in utero using fetal ultrasound

Evaluation

There is general agreement among experts that the radiologic diagnosis of CM-I in adolescents and adults is made by MRI when one or both cerebellar tonsils are displaced by ≥5 mm below the foramen magnum.

Borderline displacement of the cerebellar tonsils (≥3 to <5 mm below the foramen magnum) is considered pathologic if it is associated with additional features of CM-I, such as other craniocervical junction anomalies or syringomyelia

Management

Symptomatic patients – Decompressive surgery is indicated for patients with CM-I who are clearly symptomatic with lower cranial nerve palsies, syringomyelia, myelopathy, cerebellar symptoms, or occipital cough headache

The goals of surgery for Chiari malformations are to decompress the craniocervical junction and restore the normal flow of CSF in the region of the foramen magnum. The most common procedure is posterior decompression via suboccipital craniectomy with or without duraplasty. Other procedures include anterior decompression of the foramen magnum by odontoidectomy, and shunting.

Statistical analysis of recurrent symptoms after decompression surgery

Aim: To find the average rate of recurrent or residual syringomyelia following posterior fossa decompression as a result of Chiari malformation with associated syringomyelia.

Results

Of the 72 citations, 11 citations met inclusion criteria. Rates of recurrent/residual syringomyelia after decompression in adults range from 0 to 22% with an average of 6.7%. There were no studies that discussed specifically management of the remaining syrinx.

Inclusion criteria of patients: Adult patients with Chiari malformation and associated syringomyelia who have undergone posterior fossa decompression were included in the

study. *Exclusion criteria of patients:* Patients younger than 18 years, those sustaining trauma, meningitis, tumour, haemorrhage, arachnoiditis, and Chiari with no presence of syringomyelia, as well as those treated by any other treatment than posterior fossa decompression were excluded. Case reports, studies with fewer than 10 patients, nonhuman in vivo, in vitro, and biomechanical studies were also excluded.

Conclusions

Rates of recurrent/residual syringomyelia after posterior fossa decompression in adults range from 0 to 22% with an average across studies of 6.7%.

The information of this statistical analysis is derived from the databases included PubMed, Cochrane and National Guideline Clearinghouse databases as well as bibliographies of key articles.

LITERATURE

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EXPLORING NEURODEGENERATIVE DISEASES OF THE CENTRAL NERVOUS SYSTEM: A STUDY ON CEREBELLAR DEGENERATION TYPES AND THEIR CLINICAL IMPLICATIONS

Introduction

Neurodegenerative diseases (NDs) are a group of neurological disorders characterized by the progressive dysfunction of neurons and glial cells, leading to their structural and functional degradation in the central and/or peripheral nervous system. Neurodegenerative diseases have primarily focused on the brain, brain stem, or spinal cord associated with disease-related symptoms, often overlooking the role of the cerebellum [1].

Aging leads to the accumulation of disabilities and diseases that limit normal body functions and is a major risk factor for neurodegenerative diseases. Many neurodegenerative diseases share oxidative stress and nitrosative stress as common terminal processes. According to free radical theory of aging, an elevation in reactive oxygen species (ROS) and reactive nitrogen species (RNS) damages neural membranes and induces oxidative and nitrosative stress. The increase in oxidative and nitrosative stress is accompanied by the concomitant decline in cognitive and motor performance in the elderly population, even in the absence of neurodegenerative diseases. Markedly increased rates of oxidative and nitrosative stress are the major factors associated with the pathogenesis of neurodegenerative diseases. Diet is a key environmental factor that affects the incidence of chronic neurodegenerative diseases. Dietary supplementation with polyphenols, resveratrol, ginkgo biloba, curcumin, ferulic acid, carotenoids, flavonoids, and n-3 fatty acids exerts beneficial effects not only through the scavenging of free radicals, but also by modulating signal transduction, gene expression, and restoring optimal neuronal communication [2].