ISSN 1220-8841 (Print)
ISSN 2344-4959 (Online)

ROMANIAN
NEUROSURGERY

Vol. XXXVII | No. 2 June 2023

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DOI: 10.33962/roneuro-2023-030

Romanian Neurosurgery (2023) XXXVII (2): pp. 170-174
DOI: 10.33962/roneuro-2023-030
www.journals.lapub.co.uk/index.php/roneurosurgery

Chiari 1 malformation with Platybasia. A case report

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ABSTRACT

Background. The incidence of CM-I is estimated to be 1/1,000 births. In rare cases, CM-I is complicated by associations with other malformations of the craniocervical junction, including mainly Basilar invagination (BI). Chiari malformation (CM) is characterized by a congenital malformation of the posterior cranial fossa with cerebellar tonsils herniation through the foramen magnum, probably due to the underdevelopment of the posterior bony skeleton (exo-occipital and supraoccipital bones). CM may be complicated by a variety of other malformations, including platybasia, basilar invagination and occipitalization, although syringomyelia (SM) is the most commonly observed

Case Report. A girl, 14 years old came with a chief complaint of headache, which is exacerbated whenever the patient coughed or sneezed. The patient also complained about neck pain and a tingling sensation in her hand. A history of vomiting or seizure has not been found. A history of muscle weakness was not found. On physical examination, the patient has a GCS score of 15 on admission, with normal muscle tone and normal reflexes. The patient has a sensory deficit, on the level below C4. The patient underwent CT Scan and MRI whole spine, Chiari 1 malformation and platybasia were diagnosed. The patient underwent posterior fossa decompression, and postoperatively symptoms of Chiari and sensory deficit gradually improved.

Discussion. As an association with this syndrome, there is also basilar invagination as shown by flat basioccipt (platybasia) and upward odontoid projection. hydrocephalus [3].

Virchow coined the term 'platybasia' to describe an abnormal flattening of the skull base, a defect which he attributed to abnormal bone development. In addition to the flattening of the base of the skull, there was upward displacement (impression) of the basilar and condylar portions of the occipital bone, which caused infolding, or impression of the foramen magnum, reduction of the posterior fossa, and consequent protrusion of the upper cervical spine into the anterior brainstem, with neurological signs. The symptomatology presented by CM-1 patients is diverse, and its severity does not correlate with the degree of TH, with some asymptomatic cases presenting with prominent TH. The onset of symptoms generally develops gradually, however, trauma, coughing/sneezing or pregnancy can also precipitate the event. The most common treatment for these patients is surgical PCF decompression (alone or with duraplasty), although cerebellar tonsillectomy, cervical laminectomy, and suboccipital cranioplasty are also applied. The goal of these surgical procedures is to decompress the foramen magnum and increase the subarachnoid space in order to avoid the impaction of the cerebellar tonsils, reestablish the CSF flow and reverse the symptoms

Keywords
Chiari malformation,
Platybasia,
posterior fossa surgery

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ISSN online 2344-4959
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First published
June 2023 by
London Academic Publishing
www.lapub.co.uk

Conclusion. This patient was diagnosed with Chiari 1 malformation and platybasia, as suggested in the literature, posterior fossa decompression was done and the patient improved clinically.

Background

According to estimates, 1 in 1,000 newborns will result in CM-I. In a small percentage of instances, CM-I is complicated by relationships with other craniocervical junction abnormalities, primarily Basilar invagination (BI).[1]A cerebellar tonsil herniation through the foramen magnum is a characteristic of the Chiari malformation (CM), which is most likely caused by the posterior bony skeleton's incomplete development (exo-occipital and supraoccipital bones).[2] The cerebellar tonsils and the medial sections of the lower cerebellar lobes were pushed >5 mm past the craniospinal transition and into the upper spinal canal, resulting in the disease known as Chiari malformation type I. (CM-I).[1]

Syringomyelia (SM) is the most frequently reported abnormality, but CM may be worsened by a number of other deformities, such as platybasia, basilar invagination, and occipitalization.[3] Basilar invagination, basilar impression, and platybasia are examples of congenital and acquired abnormalities of the craniovertebral junction (CVJ), which can manifest as either abrupt or slowly advancing neurologic impairment.[4] While basilar invagination refers to the projection of the odontoid process toward the posterior fossa, platybasia refers to flattening of the skull base. Basilar impression, platybasia, brainstem kinking, and retroflexed odontoid deformities were present in 7.7% of our Chiari malformation type I patients.[6]

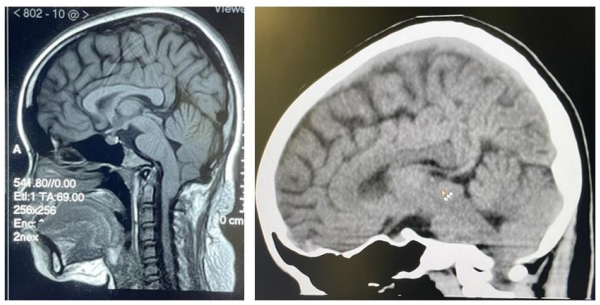

Figure 1. CT and MRI of patient, Chiari I was found.

In addition to a reduction in the size of the posterior cerebral fossa (PCF), CM is usually linked to a number of disorders including platybasia, basilar invagination, and clivus concavity. In roughly 20–72% of instances involving CSF problem, the cerebrospinal fluid (CFS) flow dynamics can deteriorate in CMI, and syringomyelia can develop most frequently in the cervical area. Nearly 20–30% of people will experience cranial-vertebral junction anomalies include basilar invagination, platybasia, a small posterior fossa, concavity of the clivus, occipitalization of the atlas, and spina bifida in the upper cervical area.[7]

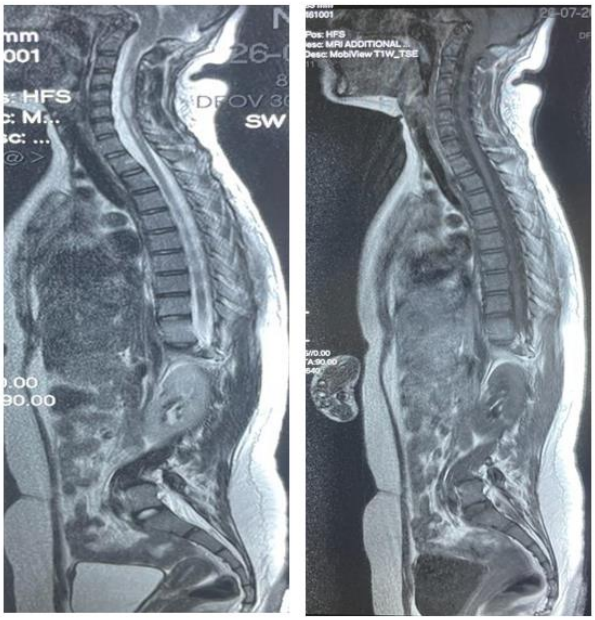

Figure 2. MRI of spine.

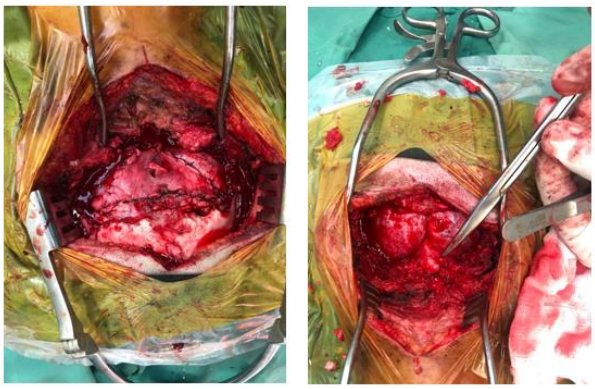

Figure 3. Posterior fossa decompression surgery done on patient.

Case Report

A girl, 14 years old came with a chief complaint of headache, which is exacerbated whenever patient

coughed or sneezed. Patient also complain about the neck pain and tingling sensation on hand. History of vomiting or seizure has not been found on patient. History of muscle weakness was not found. On physical examination, patient has GCS score of 15 on admission, with normal muscle tone and normal reflex. Patient has sensory deficit, on level below C4. Patient underwent CT Scan and MRI whole spine, Chiari 1 malformation and platybasia was diagnosed. Patient underwent posterior fossa decompression, and postoperatively symptom of chiari and sensory deficit gradually improving.

Discussions

The most common kind of "Chiari malformations," known as Chiari malformation type 1 (CM-1), is characterized by a downward herniation of the caudal section of the cerebellum into the upper cervical area through the foramen magnum.[8] Syringomyelia and a downward tonsillar herniation make up the Chiari I malformation. Additionally, basilar invagination, as evidenced by a flat basioccipt (platybasia) and an upward odontoid projection, is associated with this disease. The dens should typically extend up to 3 mm above the Chamberlain line and 5 mm above the McGregor line.[9] According to Bao et al., problems related to Chiari I malformation (CMI) included hydrocephalus in 17 patients, 51 patients with basilar impressions, 38 patients with platybasia, 32 patients with occipitalization, 67 patients with scoliosis, and 16 patients with neuropathic arthropathy.[3]

To characterize an abnormal flattening of the skull base, a deformity he linked to aberrant bone growth, Virchow developed the name "platybasia." The occipital bone's basilar and condylar portions were displaced upward, flattening the base of the skull in addition. This upward displacement resulted in reduction of the posterior fossa, infolding of the foramen magnum, and protrusion of the upper cervical spine into the anterior brainstem, along with neurological symptoms.[10] It is commonly recognized that platybasia and basilar invagination are related. In his initial investigation, Chamberlain referred to both of these things as being the same thing. Later, a number of scholars questioned platybasia's clinical value and claimed that it solely had anthropological significance. The more horizontal angulation and shortening of the clivus were related to the superior position of the odontoid process. Klaus observed that the tip of the odontoid process almost never reaches the Wackenheim clival line in basilar invagination associated with platybasia, whereas in a steeply shelving or normal clivus, the line from the dens frequently reaches or even overshadows it. The presence of platybasia suggests that it played a similar role to odontoid process invagination in producing the front concavity of the brainstem and decreasing the size of the posterior fossa.[11]

The current diagnosis of Chiari malformation type 1 relies on imaging evidence of cerebellar tonsil herniation (TH) that extends at least 3-5 mm below the foramen magnum. In other words, the resulting decreased cranial space causes overcrowding in the brain processes and causes the cerebellum to herniate through the foramen magnum. This directly compresses the neural tissue at the craniovertebral junction and frequently causes disruptions in the cerebrospinal fluid (CSF) (decreased velocity and increased impedance), which can lead to various related diseases. The symptoms displayed by CM-1 patients are varied, and the severity of the symptoms does not always correspond to the degree of TH, with some asymptomatic instances exhibiting substantial TH. The onset of symptoms often happens gradually, although it's possible for trauma, coughing/sneezing, or pregnancy to trigger it.[8] For more than a century, basilar invagination has been associated with physical characteristics such a short neck, low hairline, web-shaped neck muscles, torticollis, a reduction in the range of neck movements, and several other morphological abnormalities. There have also been reports of other bone fusion abnormalities and platybasia. It is common to experience neck pain, muscle spasms, and restricted neck movement, all of which point to localized instability.[11]

The most effective diagnostic approach for identifying Chiari malformation is MRI. MRI can reveal precise anatomical details on the structures in the cranial basement. The diameter of the foramen magnum, the volume of the PCF, and the condition of the platybasia were measured using cranial distance and angle measurements. Measurements of PCF development and platybasia were made using the slope of the Tentorium Cerebelli. The patient group showed a considerable reduction in distances measured for the evaluation of platybasia, such as the distance between the Chamberlain line and the tip of the dens axis, the Klaus index, the clivus length,

and the distance between the internal occipital protuberance and the opisthion. The measurement of head basis angles reveals a platybasia tendency.[7]

Although cerebellar tonsillectomy, cervical laminectomy, and suboccipital cranioplasty are also used, surgical PCF decompression (either alone or in conjunction with duraplasty) is the most often used treatment for these patients. In order to prevent the impaction of the cerebellar tonsils, restore the CSF flow, and treat the symptoms, these surgical treatments aim to widen the foramen magnum and enhance the subarachnoid space.[8] To choose the best care and compare surgical results, it is becoming increasingly important to divide all other cases into particular subgroups. Chiari malformation type I is the general term for the results of CMI patients who have a platybasia or a hypoplasic posterior cerebral fossa. Actually, the same surgical procedure, posterior fossa decompression, with or without dural expansion, might result in a further hindbrain descent in the first case or in "ascent" of the cerebellar tonsils in the second case.[12] Marin-Padilla concluded from their study that the Chiari-like deformities reflect the effects of clival and occipital molding, which act mainly anteriorly. Platybasia did not directly result in any neurological symptoms, but it participated with basilar invagination in critically reducing the posterior cranial fossa volume.[11]

Conclusion

Chiari malformation is generally related to occipital bone dysplasia and it is frequently associated with various conditions such as platybasia, basillar invagination and clivus concavity in addition to decrement in posterior cranial fossa (PCF) size. Chiari malformation type 1 results in a direct compression of the neural tissue at the craniovertebral junction and, often, cerebrospinal fluid (CSF) disturbances (decreased velocity and elevated impedance), that can cause other related conditions and bone fusion deformities such as platybasia. MRI method is the best diagnostic tool for detecting Chiari malformation. MRI provides detailed anatomical information regarding the structures located at the cranial basement. The most common treatment for these patients is surgical PCF decompression (alone or with duraplasty), although cerebellar tonsillectomy, cervical laminectomy, suboccipital cranioplasty are also applied. The goal of these surgical procedures is to decompress the foramen magnum and increase the subarachnoid space in order to avoid the impaction of the cerebellar tonsils, reestablish the CSF flow and reverse the symptoms.

List of Abbreviation

CM: Chiari Malformation
CSF: Cerebrospinal fluid
CT: Computed Tomography
CVJ: Craniovertebral Junction
GCS: Glasgow Coma Scale
MRI: Magnetic Resonance Imagin
PCF: Posterior Cranial Fossa
SM: syringomyelia
TH: Tonsillar herniation

References

1. Brito JNP; Santos BA; Nascimento IF; Martins LA; Tavares CB. Basilar invagination associated with chiari malformation type I: A literature review. Clinics 2019;74:653.
2. Joaquim AF. Severe Cerebellar Degeneration and Chiari I Malformation – Speculative pathophysiology based on a systematic review. Rev Assoc Med Bras. 2020; 66(3): 375-379.
3. Bao C; Yang F; Liu L; Wang B; Li D; Gu Y; et al. Surgical treatment of Chiari I malformation complicated with syringomyelia. Experimental and Therapeutic Medicine. 2013; 5: 333-337.
4. Pinter, N.K., McVige, J. & Mechtler, L. Basilar Invagination, Basilar Impression, and Platybasia: Clinical and Imaging Aspects. Curr Pain Headache Rep 20, 49 (2016). Available from: https://link.springer.com/article/10.1007%2Fs11916-016-0580-x [Accessed on 16th September 2021]
5. Silva ATPB, Silva LTPB, Vieira AENR, Melo CIE, Nascimento JJC, Mello Júnior CF, Vasconcelos SC, Araújo-Neto SA. Craniometric parameters for the evaluation of platybasia and basilar invagination on magnetic resonance imaging: a reproducibility study. Radiol Bras. 2020 Set/Out;53(5):314–319
6. Royo-Salvador MB; Fiallos-Rivera M; Salca H. Caudal Traction as a Pathogenetic Mechanism of Chiari Malformation Type I. IntechOpen. 2019. Available from: https://www.intechopen.com/chapters/70103 [Accessed on 16th September 2021]
7. Alkoc OA; Songur A; Eser O; Toktas M; Gonul Y; Esi E; et al. Stereological and Morphometric Analysis of MRI Chiari Malformation Type-1. Journal of Korean Neurosurgical Society 2015; 58(5): 454-461.
8. Urbizu A, Khan TN, Ashley-Koch AE. Genetic dissection of Chiari malformation type 1 using endophenotypes and stratification. J Rare Dis Res Treat. (2017) 2(2): 35-42
9. Abdrabou, A. Chiari 1 malformation with basilar invagination. Case study, Radiopaedia.org. (accessed on

16 Sep 2021) https://radiopaedia.org/cases/34375
10. Pearce JMS. Platybasia and Basilar Invagination. Eur Neurol 2007;58:62–64
11. Goel A. Basilar invagination, Chiari malformation, syringomyelia: A review. Neurol India 2009;57:235-46
12. Rocco CD. Should we stop using the term “malformation” for Chiari type 1?. Child's Nervous System (2019) 35:1649–1650.