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Morphometric analysis of posterior fossa and craniovertebral junction in subtypes of Chiari malformation

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ABSTRACT

Objectives: Chiari malformations (CMs) are a group of disorders defined by anatomic anomalies of the cerebellum, brainstem, and craniovertebral junction (CVJ). In this study, we aimed to investigate morphometry of posterior fossa and CVJ in subtypes of CM and in control group, and to bring up a matter a correlation with demographic data and subtypes of CM.

Patients and Methods: The study group included patients managed for CM between 2012 and 2016 and control group. Radiological evaluation was studied by special programs and formulas. Intracranial volumes and morphometric datas of posterior fossa and CVJ were recorded retrospectively.

Results: Of the 141 patients, 91 had CM and 50 were control group participants. Mean age was 34.75. Patients were classified as CM-0 (n:10), CM-1 (n:45), CM-1.5 (n:21), CM-2 (n:15). There were statistically significance between Chiari subtypes by syringomyelia (SM) presence ($p < 0.01$), SM localization ($p < 0.01$), posterior fossa volume (PFV) ($p < 0.01$), length of clivus (LoC) and length of suboccipital (LoSO) ($p < 0.01$ for both), angle between clivus and suboccipital (C-SO angle) ($p < 0.01$), and clivo-dental angle (C–D angle) ($p < 0.01$).

Conclusion: On morphometric comparison of CM subtypes we concluded that etiological differences lead to morphological differences. CM-2 has remarkable differences from both other subtypes and the control group.

1. Introduction

Chiari malformations (CMs) are a group of disorders defined by anatomic anomalies of the cerebellum, brainstem, and craniovertebral junction (CVJ), characterized by downward displacement of the cerebellum into the spinal canal, either alone or together with the lower medulla [1]. There is no certain definition for amount of cerebellar descent. CMs were first described in 1883 by John Cleland [2,3]. In 1891, Hans Chiari classified CMs into four groups [4]; today, they are divided into six groups as Chiari 0, 1, 1.5, 2, 3, and 4 [4–7]. Beyond the earlier classification, Chiari 0 malformation (CM-0) was described as syringomyelia (SM) despite the lack of cerebellar tonsil herniation [8], and Chiari 1.5 malformation (CM-1.5) as a tonsillar herniation within a Chiari I malformation (CM-1), with additional caudal descent of the brainstem through the foramen magnum [5,7]. It is thought that Chiari 1 malformation disorders arise from para-axial mesoderm [9]. Although, Chiari 2, 3 and 4 arise from neuroectoderm [10]. The components of the posterior fossa outgrow the underdeveloped compartment and cause herniation of the tonsils into the upper cervical spinal canal [11]. A number of studies have attributed this insufficient posterior cranial fossa geometry to embryological defects in the paraxial mesoderm [12–14].

For accurate diagnosis and treatment of various diseases, it is important for clinicians to know the normal anatomy of the cranial base, especially the foramen magnum and associated structures [15]. As the cranial base can remain intact in cases where the rest of the cranium has been compromised, researchers have analyzed dimorphic traits of this anatomic region for the purposes of gender identification [16,17].

In this study, our objectives are 1) to establish whether intracranial volumes differ in individuals with various subtypes of CMs as compared to healthy individuals; 2) to investigate the correlation within intracranial volumes and CVJ morphometries; 3) to investigate the correlation between syringomyelia and posterior fossa morphometry; 4) to investigate the correlation between CM etiology and posterior fossa morphometry.

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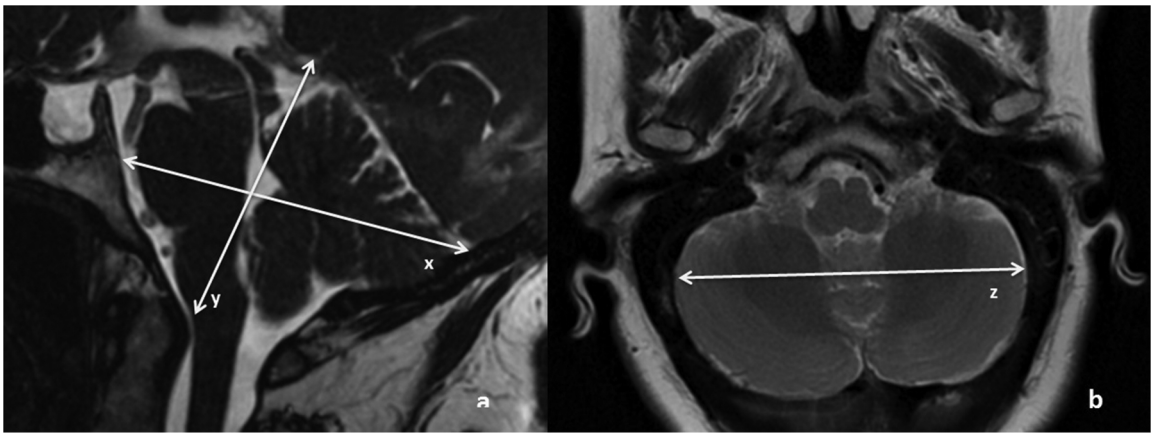

Fig. 1. Spheroidal PFV was calculated using the simple spheroidal formula.

2. Material and methods

2.1. Patient population

Following a retrospective review of patients evaluated or operated for CM at the hospital between December 2012 and February 2016, 91 patients and 50 control cases were included in the study. The study group included patients with or without all CM symptoms, excluding those for whom the available data were insufficient to evaluate cranial volume and morphology, we excluded patients with intracranial pathology and patients with CM-3 because of the bone defect they have. Control cases included patients admitted to the hospital for headache or for reasons other than intracranial pathology.

2.2. Definition of CM

Subtypes of CMs are almost completely different diseases with diversified clinical and radiological features. According to the previous classification [1,10],

- Chiari I malformation (CM-I) is characterized by abnormally shaped cerebellar tonsils displaced below the level of the foramen magnum;
- Chiari II malformation (CM-II) is characterized by downward displacement of the cerebellar vermis and tonsils, brainstem malformation with beaked midbrain on neuroimaging, and spinal myelomeningocele;
- Chiari III malformation (CM-III) is rare and combines a small posterior fossa with a high cervical or occipital encephalocele, usually with displacement of cerebellar structures into the encephalocele, and often with inferior displacement of the brainstem into the spinal canal;
- Chiari IV malformation (CM-IV) is now considered to be an obsolete term that describes cerebellar hypoplasia unrelated to the other CMs.

In addition to the previous classification,

- Chiari 0 malformation is SM despite the lack of cerebellar tonsil herniation [8];
- Chiari 1.5 malformation involves tonsillar herniation within a Chiari I malformation, with additional caudal descent of the brainstem through the foramen magnum [7].

2.3. Radiological evaluation

All radiological values were reported from a single measurement from the same observers (RB). A lateral view radiograph of the CVJ was used to evaluate CMs. All patients underwent magnetic resonance imaging (MRI) (Signa 1.5-Tesla; General Electric), using T2-weighted MRI sequence for all measurements. Linear dimensions were derived using ExtremePacs Workstation 1.5 software (Extreme PACS Healthcare, Ankara, Turkey). To determine a plane parallel to the foramen magnum (FM), MRI of the CVJ was performed at 5 mm intervals parallel to the orbitomeatalline. Measurement of cerebellar descent (CD) was performed on MRI sequences using the same software. Examination protocol is slices parallel to the line joining the Genu and Splenium of the Corpus Callosum. Standart setting are 4–5 mm of slices, 0.9 mm of resolution, 10–40% of gap and 230–250 mm of FOV.

2.4. Measurement of volume

Spheroidal PFV was calculated using the simple spheroidal formula [18]

$$PFV = 4/3 \times \Pi \times (X/2 \times Y/2 \times Z/2),$$

Where x is the anteroposterior measurement from the posterior clinoid process to the torcula; y is the height of the posterior fossa measured from the basion to the peak of the tentorium cerebelli; and z is the maximum width of the posterior fossa (Fig. 1).

In children, ICV was calculated using the Dekaban spheroidal formula, which estimates cranial volume in individuals up to 20 years of age[19–21]:

$$ICV\ (cm^3) = 0.523 \times (length - 2\,t) \times (breadth - 2\,t) \times (height - t),$$

Where t = thickness of skull and scalp.

In adult males and females, ICV was calculated using the formula derived by Lee-Pearson [20,21]:

$$Adult\ male\ ICV\ (cm^3) = [0.000337 \times (length - 11).(breadth - 11).(height - 11) + 406.01\ cm^3;$$

$$Adult\ female\ ICV\ (cm^3) = [0.0004 \times (length - 11).(breadth - 11).(height - 11) + 206.60\ cm^3.$$

In adults, maximum anteroposterior (AP) length was measured between the glabella and the inion; maximum breadth was measured between the two parietal eminences; and cranial height was measured as basibregmatic height (Fig. 2).

2.5. Measurement of foramen magnum area

The area of the foramen magnum (FMA) was calculated using the formula derived by Radinsky [22]:

$$FMA = 1/4 \times FML \times FMW,$$