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N. Boggio et al., Bihemispheric PICA with Chiari I

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Abstract
Typically, patients with Chiari I malformations (CM I) do not have other intracranial anatomical variations, especially vascular derailments. Here, we report the findings of a cadaveric specimen found to have CM I and cerebellar tonsils supplied by a single posterior inferior cerebellar artery (PICA) i.e., a bihemispheric PICA. An adult male cadaver was found to have CM I. It was also noted that the left PICA descended inferiorly to the level of C1 and that there was absence of the
right PICA. The territory of the right PICA was supplied by the left PICA. The tonsillar component of the left PICA gave rise to a branch that crossed to the right inferior cerebellum and herniated cerebellar tonsil. A bihemispheric PICA is very rare. To our knowledge, this is the first report of this vascular variation in combination with CM I. Such a variation should be kept in mind, especially during posterior fossa decompression for symptomatic CM I as unilateral PICA injury could have catastrophic results.

Keywords: Hindbrain herniation, vertebrobasilar system, posterior cranial fossa, tonsillar ectopia, variation

INTRODUCTION

Chiari malformations are congenital hindbrain anomalies originally described during the 1890s by Hans Chiari [13,14]. While Chiari’s traditional classification system comprised four types of malformations, more recent and specific systems recognize eight classes: Chiari 0, Chiari I, Chiari 1.5, Chiari II, Chiari III, Chiari 3.5, Chiari IV, and Chiari V [8,42]. Chiari type I malformation (CM I) involves a caudal herniation of the cerebellar tonsils greater than 5 mm inferior to the plane of the foramen magnum [1,5,40]. In 2002, Tubbs et al. found that 96% of CM I patients exhibit asymmetric tonsillar herniation. The herniated tonsils often undergo chronic compression, often succeeded by atrophy and the loss of their folia, which can make them smooth in appearance [12]. However, Cesmebasi et al. noted that apart from the herniated cerebellar tonsils, the brain of a CM I patient is generally normal.

The most common symptom of CM I is pain, occurring in 60-70% of patients, often in the occipital or upper cervical regions [29]. Exacerbated by Valsalva maneuvers such as laughing, coughing, or sneezing, Chiari-like headaches, which are posteriorly located and short in duration, have also been described [4]. Tubbs et al. (2004) and Cesmebasi et al. (2015) reviewed associated anatomical findings often seen in CM I patients involving the skull, spine, meninges, and spinal cord [12,40]. Common skull-related findings include occipital bone dysplasia, and basilar skull and cranio cervical junction anomalies such as a large foramen magnum, a short clivus, and a shallow posterior cranial fossa [26,28,35,38,39,43]. Spinal symptoms commonly associated with CM I include the Klippel-Feil deformity; atlantoaxial assimilation; and scoliosis, often a levoscoliosis [29,37,43]. More specifically, the incidence of
scoliosis in CM I patients could be as high as 30%, and up to 70% if syringomyelia is also present [32]. If it is caused by syringomyelia, levoscoliosis of the thoracic vertebrae can present as abnormal abdominal reflexes, desensitization to pain and temperature, or non-specific, non-dermatomal flank or back pain [12]. CM I patients often exhibit symptoms in the meninges such as an elevated slope of the tentorium cerebelli, thickening of the arachnoid mater at the level of the foramen magnum, and/or arachnoid veils obstructing the outlets of the fourth ventricle [12]. Among CM I patients, 50-75% present with a syrinx, usually located in the upper thoracic or lower cervical spine [3,7,9,11,12,26]. Other symptoms commonly associated with CM I include weakness or numbness, unsteadiness, ophthalmological or otological disturbances, atrophy, hyperreflexia, ataxia, and lower cranial nerve dysfunction [29].

Currently, decompression surgery of the posterior cranial fossa is widely accepted as the only effective treatment option for patients with symptomatic CM I [7,12]. The aim of the surgery is to alleviate symptoms and inhibit progressive deterioration by enlarging the posterior fossa region and restoring CSF flow from the fourth ventricle to the cervical subarachnoid space [29].

Here, we report the findings of a cadaveric specimen found to have CM I and cerebellar tonsils supplied by a single posterior inferior cerebellar artery (PICA).

**CASE REPORT**

During the routine dissection of the posterior cranial fossa and upper cervical spine in a 89-year-old at death male cadaver, a CM I was identified (Fig. 1). The left cerebellar tonsil was 5 mm inferior to the foramen magnum and the right cerebellar tonsil was 10 mm inferior to the foramen magnum at the level of the C2 nerves rootlets. It was also noted that the left PICA descended inferiorly to the level of C1 and that there was absence of the right PICA. The territory of the right PICA was supplied by the left PICA (Fig. 1). The left PICA arose from the V4 segment of the vertebral artery. The vessel took a normal course by the medulla oblongata and as mentioned above, its tonsillar segment, which hugged the inferior aspect of the left herniated cerebellar tonsil was descended to the level of C1. Small branches supplied the left cerebellar tonsil and inferior surface of the cerebellum. The tonsillar component of the left PICA then, at the midline, gave rise to a branch that crossed to the right inferior cerebellum and
herniated cerebellar tonsil although this branch did not course around the caudal pole of the tonsil but over its posterior upper surface (Fig. 1). The posterior spinal artery was contributed to by the left PICA and a contralateral branch of this vessel deep to the right cerebellar tonsil. No other vascular or other intracranial anatomical variations were noted. No intracranial pathology such as hemorrhage, obvious ischemic changes, or hydrocephalus were found. The cause of death in the cadaver donor was myocardial infarction.

DISCUSSION

The arterial supply to the cerebellum typically involves three sets of paired arteries arising from the vertebrobasilar system: the superior cerebellar artery (SCA), the anterior inferior cerebellar artery (AICA), and the PICA. The PICA is most relevant to a discussion of cerebellar tonsil herniation. Along its tortuous course, this artery supplies the lower medulla oblongata, choroid plexus, dura of the posterior cranial fossa, fourth ventricle, cerebellar tonsils, vermis, and inferolateral hemisphere. The PICA most commonly originates as the largest branch of the bilateral intracranial vertebral arteries, often near the vertebrobasilar junction. Although its origin and course are highly variable, the PICA trunk is often divided into five segments as it travels inferiorly toward the foramen magnum: anterior medullary, lateral medullary, tonsillomedullary, telovelotonsillar, and cortical (Fig. 2) [23,17,27,33,41]. These authors, among others, described those segments as follows. The initial segment, the anterior medullary segment (1), starts at the origin of the PICA, courses by the anterior medulla, and ends at the medial edge of the inferior olive. From there, the lateral medullary segment (2) extends towards the origins of the lower cranial nerves (CNs IX, X, and XI) at the lateral edge of the inferior olive. Continuing as the tonsillomedullary (posterior medullary) segment (3), the PICA descends to the inferior pole of the cerebellar tonsil and then ascends to the midpoint of the tonsil on its medial surface, forming its caudal loop. The telovelotonsillar (supratonsillar) segment (4) begins at the midportion of the tonsil and continues the ascent of the PICA to the roof of the fourth ventricle before it recurs to form the cranial loop, which contains the choroidal point, then descends posteriorly to the fissures between the tonsil, vermis, and cerebellar hemisphere. The final segment (5), the cortical segment, arises from the tonsillobiventral fissure and often bifurcates in its course around the
tonsil into a medial (vermian) trunk, giving off vermician branches, and a lateral (tonsillohemispheric) trunk, giving off tonsillar and hemispheric branches [23].

**Major variants of the posterior inferior cerebellar artery**

As mentioned, the origin and course of the PICA often vary. Miao et al. (2020) reviewed the literature thoroughly and detailed the variations of the PICA as follows. Although this artery most commonly originates from the intradural vertebral artery near the vertebrobasilar junction, its origin has been observed at all points along the intracranial vertebral artery and from every direction [2, 23]. Additional variations in the location of its origin include: the extradural vertebral artery (0.4-20.8% of cases) [23,24,30,31], the basilar artery (6-11% of cases) [16], an AICA-PICA common trunk off the basilar artery, and, less commonly, from other arteries such as the internal carotid, primitive hypoglossal, primitive trigeminal, and posterior meningeal [27]. Several variations relating to the manner in which the PICA arises and its subsequent course have also been described. For example, in 2.8-7% of reported cases, the vertebral artery continued as the [31]. A duplicated origin of the PICA (two distinct arteries without convergence) has been observed in 2-6% of hemispheres [16,23,36], and a double origin (two distinct origins with distal convergence) in approximately 1.45% of cases [22]. A fenestrated PICA is very rare, occurring in only about 0.3% of cases (Pekcevik, 2014). A hypoplastic PICA has a reported prevalence of 15-32% [16,20]. Significantly, the PICA has been reported unilaterally absent in 6-26% of cases and bilaterally absent in 2-3.6% [6, 19,23,31,34,44].

The distal portion of the PICA is also very variable. While its cortical segment most commonly bifurcates into a medial (vermian) and a lateral (tonsillohemispheric) trunk as it courses over the posterior surface of the tonsil, the locations of both the bifurcation and the division of branches can vary [23,25]. For example, the tonsillar branches can arise from the medial trunk with the vermian branches, rather than from the lateral trunk [41]. Lister et al. (1982) noted “a reciprocal relationship with frequent overlap in the areas supplied by the tonsillar, hemispheric, and vermian branches” (p. 193) [23]. The tonsillar branches were most often observed suppling the medial, posterior, inferior, and a portion of the anterior surface of the tonsil. The number of tonsillar branches arising from a single PICA ranged from zero to five (average 1.6), with six out of 50 cerebellar hemispheres having zero tonsillar branches. In such cases, the adjacent hemispheric and vermian branches supplied the cerebellar tonsils [23].
**Posterior Inferior Cerebellar Artery and Chiari I Malformation**

In some instances, such as cases of hindbrain herniation seen in CM I, the PICA extends extracranially as seen in the present case report [41]. Margolis and Newton (1971) posited that despite significant variation in the sites at which the lateral trunk gives rise to tonsillar and hemispheric branches, it is because of their common origin that the branches are affected by hindbrain herniation [25]. Moreover, in CM I, it is the caudal herniation of the cerebellar tonsils that pulls on the lateral trunk and ultimately stretches both the tonsillar and hemispheric branches inferiorly. Angiographic visualization of these caudally displaced branches below the level of the foramen magnum has previously assisted in the diagnosis of Chiari malformations, particularly Chiari II malformation, in which the cerebellar vermis, brainstem, and fourth ventricle are also displaced through the foramen magnum [18,42]. Some authors have described a stretched-out, hairpin caudal loop of the PICA below the level of the foramen magnum associated with CM I, though the diagnostic value of this has been questioned owing to the immense variability of the course of the PICA [25,18].

**Bihemispheric PICA**

Upon further analysis of the AICA and PICA blood supply, Fujii et al. (1980) posited that there is an inverse relationship between these two arteries with respect to both their diameters and the areas they supply [16]. Furthermore, in cases where one PICA is hypoplastic or absent, the ipsilateral AICA or the contralateral PICA is larger and supplies the area that it would normally have supplied. The authors noted an example in which the left PICA was hypoplastic and the large right PICA traveled across the midline to give rise to the left tonsillohemispheric trunk and send choroidal branches to the choroid plexus. This “extensive type” of PICA, which forms an arterial bridge as it crosses the midline to give branches to the other side of the vermis and occasionally extends over the opposite hemisphere, was observed in 14% of the 50 adult cadaveric cerebellar hemispheres that Fujii et al. examined. Cullen et al. (2005) described a very similar PICA variation in which, when one PICA was hypoplastic or absent, as in our case, the contralateral PICA crossed the midline to supply both cerebellar hemispheres; they termed this a “bihemispheric PICA” [15]. The authors detailed four cases of a bihemispheric PICA, each of which originated from the dominant vertebral artery and gave rise to vermian branches distal to
the choroidal point, and then crossed the midline along the dorsal aspect of the vermis and extended onwards to supply the contralateral hemisphere. Cullen et al. posited two subtypes, the true bihemispheric PICA, supplying both cerebellar hemispheres from a single trunk, and the vermian variant, providing the only bilateral supply to the vermis from a single trunk. Three potential mechanisms of development of these types of variations are: a midline bridging of a pial network of vessels; the development of a midline structure, such as the vermis, which could influence a vessel to cross; or fusion with a midline structure such as the basilar artery [10,15]. In 2005, Cullen et al. suggested the bihemispheric PICA to be extremely rare, less than 0.1%, but in 2013 they proposed a much higher prevalence, closer to 3%. Upon examination of 11 cases of bihemispheric PICAs, Carlson et al. observed true bihemispheric PICAs to be four times more common than the vermian variant. Each of the true bihemispheric PICAs were observed in conjunction with contralateral PICA aplasia, whereas the vermian variants were found with a normal contralateral PICA. Regardless of the subtype, all 11 cases involved the PICA crossing the midline near its cortical segment bifurcation into medial and lateral trunks. It is important to note that a true bihemispheric PICA is different from a ‘PICA communicating artery’ in that no anastomotic connections are formed, as the contralateral PICA is aplastic. Since the incidence is higher than previously recognized, the possibility of a bihemispheric PICA in patients is of clinical relevance. This is particularly true in instances of posterior cranial fossa cerebrovascular disease, such as cerebellar arteriovenous malformation, aneurysm, and stroke and ischemic syndromes. If the problem involves a bihemispheric PICA, the risk of complications such as bilateral cerebellar infarction must be considered [10,15].

CONCLUSIONS

A bihemispheric PICA is very rare. To our knowledge, this is the first report of this vascular variation in combination with CM I. Such a variation should be kept in mind, especially during posterior fossa decompression for symptomatic CM I as unilateral PICA injury could have catastrophic results.

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REFERENCES


**Figure 1.** Posterior view of the exposed craniocervical junction in the case described herein. Note the herniated left and right cerebellar tonsils (T), C2 dorsal nerve roots, posterior spinal artery (PSA), and left spinal accessory nerve (CNXI). The left PICA is seen at the *. The contralateral branch of the left PICA is seen at the arrows.

**Figure 2.** Schematic drawing of the normal course of the left PICA.