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MR Demonstration of Ectopic Fourth Ventricular Choroid Plexus in Chiari II Malformation

James E. Stark¹ and Charles M. Glasier¹

Summary: An enhancing nodule was seen on T1-weighted enhanced scans at the tip of the caudally displaced cerebellar vermis in three patients with Chiari II malformation. The enhancing fibrovascular nodule found at the base of the cerebellum in patients with Chiari II malformation represents ectopic choroid plexus and should not be confused with a pathologic mass.

Index terms: Chiari malformations; Choroid plexus, magnetic resonance; Choroid plexus, abnormalities and anomalies; Spina bifida; Pediatric neuroradiology

The magnetic resonance (MR) appearance of the hindbrain deformity characteristic of the Chiari II malformation has been well documented (1). The cerebellum, with transverse growth constrained by a small posterior fossa, grows in a vertical fashion. The superior vermis extends high up between the posterior cerebral hemispheres while the inferior vermis is located in the cervical or even upper thoracic spinal canal (2, 3). Surgeons and pathologists have noted the presence of fibrovascular tissue containing choroid plexus located between the distal tip of the vermis and medulla (3, 4). We have noted in three patients that this tissue, similar to choroid plexus in orthotopic locations, enhances intensely after the administration of intravenous Gd-DTPA and should not be confused with a pathologic mass.

Patients and Methods

Three children with known spina bifida aperta underwent spinal MR examination because of suspected syringohydromyelia or tethering of the spinal cord. The patients ranged in age from 2 to 11 years. MR scans were performed on a 1.5-T system. Sagittal and axial T1-weighted scans were obtained both before and after the administration of intravenous Gd-DTPA (0.1 mmol/kg). Axial T2*-weighted gradient-echo scans were obtained before the administration of intravenous contrast. In two patients otherwise

typical for Chiari II malformation, intraspinal neurenteric and neuroectodermal cysts were surgically excised from the cervical and thoracic spine. In the third patient, a cervical syrinx was shunted. Posterior fossa exploration was not performed in any of these three patients.

Results

All three patients showed enhancing nodular tissue at the tip of the caudally displaced cerebellar vermis on postcontrast T1-weighted images. This tissue was isointense to cerebellum on the precontrast images (see Figs. 2 and 3).

Discussion

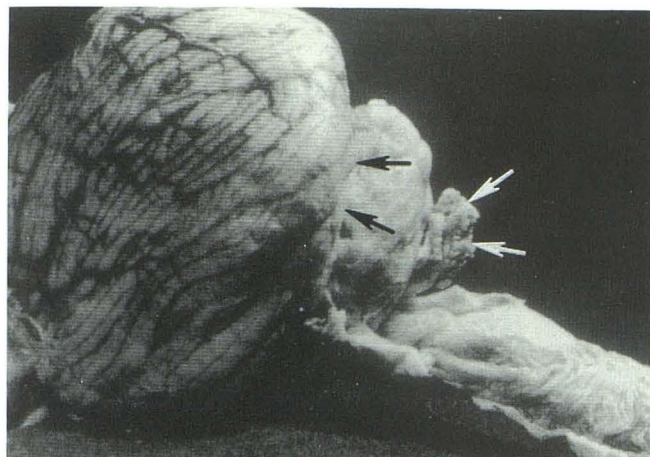
A unified theory to explain the embryologic origin and the gross pathologic abnormalities found in patients with spina bifida aperta and Chiari II malformation has recently been proposed. The initial insult in spina bifida, which occurs during the fourth week of development, is the failure of normal closure of the neural tube at the level of the posterior neuropore, leading to leakage of cerebrospinal fluid from the central canal of the spinal cord at the time that the intracerebral and intracerebellar vesicles are forming. Collapse of the cerebellar vesicles leads to decreased inductive effects on the mesenchyme of the posterior fossa and results in a small posterior fossa. During the fifth week, the pontine flexure fails to form, and the growing hindbrain is then forced to grow in a vertical fashion, superiorly through a dysplastic falx and caudally into the foramen magnum and spinal canal (5).

Extension of the inferior cerebellar vermis into the spinal canal usually accompanies the caudal

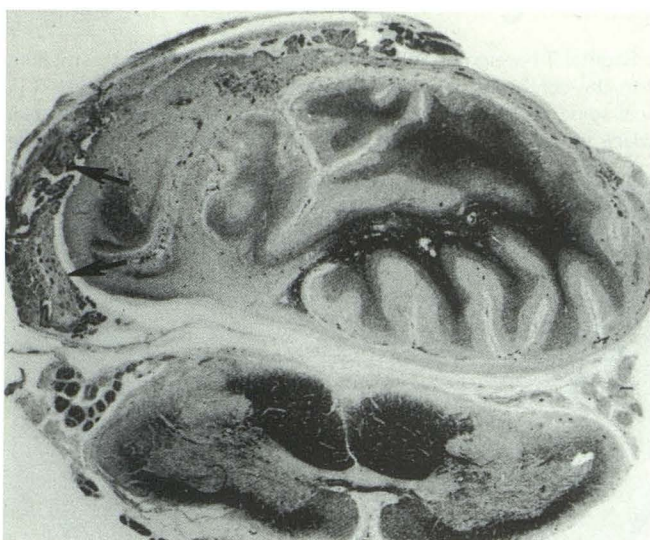
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A



B

Fig. 1. A, Gross pathologic specimen of hindbrain and cervical cord from patient with Chiari II malformation. Note nodular tuft of choroid plexus protruding beneath the cerebellar tail (*white arrows*). Impression made on the herniated hindbrain by the foramen magnum is easily seen (*black arrows*).

B, Transverse section through the cerebellar tail and cervical spinal cord shows choroid plexus and fibrous tissue (*black arrows*) dorsal and lateral to the cerebellar tissue which is covered by ependyma. (From Daniel and Strich (3).)

displacement of the medulla and pons in Chiari II malformation. This may or may not be accompanied by herniation of the cerebellar tonsils. In 2% of cases, only the cerebellar tonsils protrude caudally (2, 6). The distal tip of the vermis may lie at any level between the first cervical and second thoracic vertebral body. In a minority of cases, 25% in one series (7), the tip may even extend below the level of the medullary kink.

In Chiari II malformation, the inferior medullary velum is usually absent and choroid plexus is only rarely present within the fourth ventricle except in those patients who have cystic prolongation of the fourth ventricle and absent foramen of Magendie (2, 5, 8). Instead, a nodular fibrovascular mass containing choroid plexus tissue is usually present protruding between the vermis and medullary kink (Figs. 1A and 1B). This tissue may lie in various positions including the junction of the vermician "peg" and medulla, extending from the distal vermis to the medullary kink or on the dorsal surface of the vermis (2, 9). This tissue appears both on gross pathologic examination and on sagittal MR as a "cluster of grapes." The fibrovascular tissue is often seen on noncontrast images and has MR signal characteristics similar to those on the adjacent vermis. Occasionally, the ectopic choroidal tissue may have increased signal relative to the cerebellar vermis when ischemia secondary to compression or vascular compromise at the foramen magnum causes decrease in the vermician signal intensity on precontrast T1-weighted scans (1). Intense enhancement is noted on T1-weighted images after the administration of intravenous contrast material (Figs. 2 and 3). Intravenous contrast is not routinely used in Chiari II patients, but was administered in two patients because of the presence of surgically proved neurenteric and neuroectodermal cysts and in the third patient because of unusual, nodular-appearing neural tissue in the caudal thecal sac.

This aberrant location of fourth ventricular choroid plexus tissue apparently has its origin in early fetal life. In the normal 6-week human embryo, a portion of the cerebellum is intraventricular with the tuft of fourth ventricular choroid plexus located caudally with respect to the cerebellar tissue (Fig. 4). In patients with Chiari II malformation, the constricted fetal posterior fossa prevents formation of the normal pontine flexure and the intraventricular portion of the cerebellum enlarges and herniates inferiorly, pushing before it the choroid plexus which comes to lie dorsal and caudal to the low-lying vermis (8, 10, 11).

Shortening of T1 relaxation time with increased signal intensity on postcontrast T1-weighted images occurs normally in tissues with an absent blood-brain barrier, including choroid plexus, pineal gland, and both the anterior pituitary gland and infundibulum. Enhancement of the fibrovascular nodule found at the inferior border of the

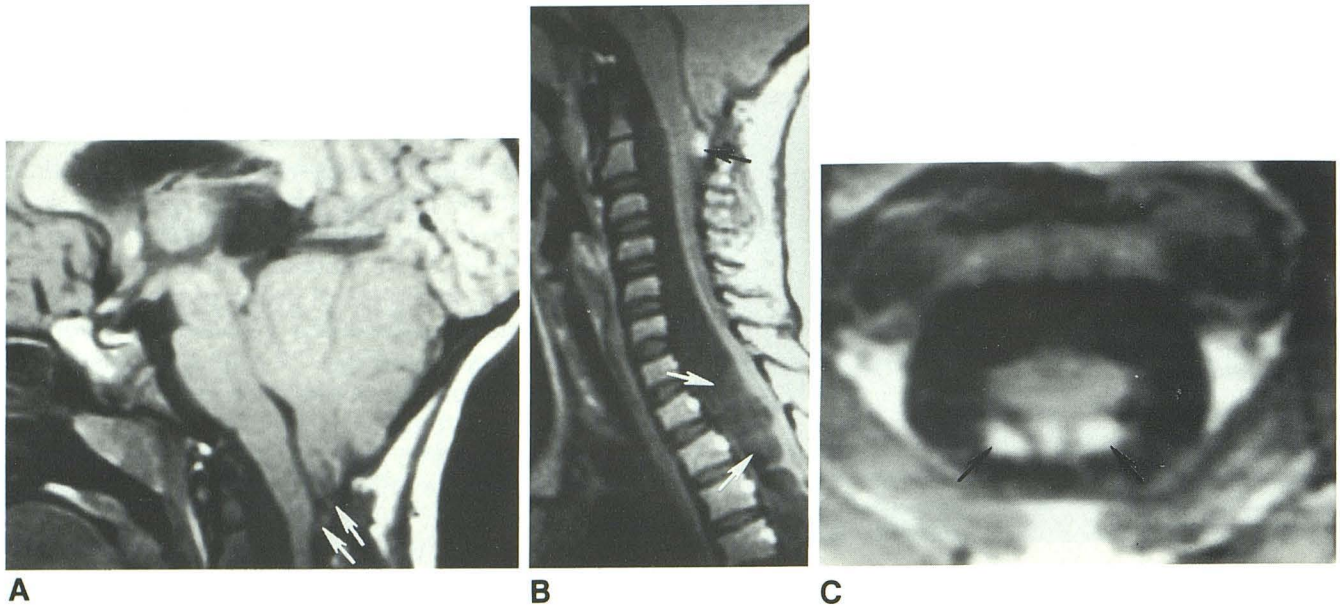


Fig. 2. Five-year-old boy with spina bifida and periscapular pain. A, Sagittal T1-weighted 600/20 (TR/TE) scan through the midline of the posterior fossa shows nodular tissue isointense to brain caudal to the cerebellar tail (*white arrows*). B, Contrast-enhanced T1-weighted sagittal 900/20 and C, axial 800/25 scans through the cervical spine show intense contrast enhancement of choroid plexus caudal and lateral to the cerebellar tail and dorsal to the cervical cord (*black arrows*, B and C). Surgically proved neurenteric cyst (*white arrows*, B).



Fig. 3. Eleven-year-old girl with spina bifida underwent spinal MR prior to scoliosis surgery. Sagittal T1-weighted 800/25 contrast-enhanced MR shows an enhancing nodule of ectopic choroid plexus just below the cerebellar tail (*arrow*). Also note midcervical syrinx.

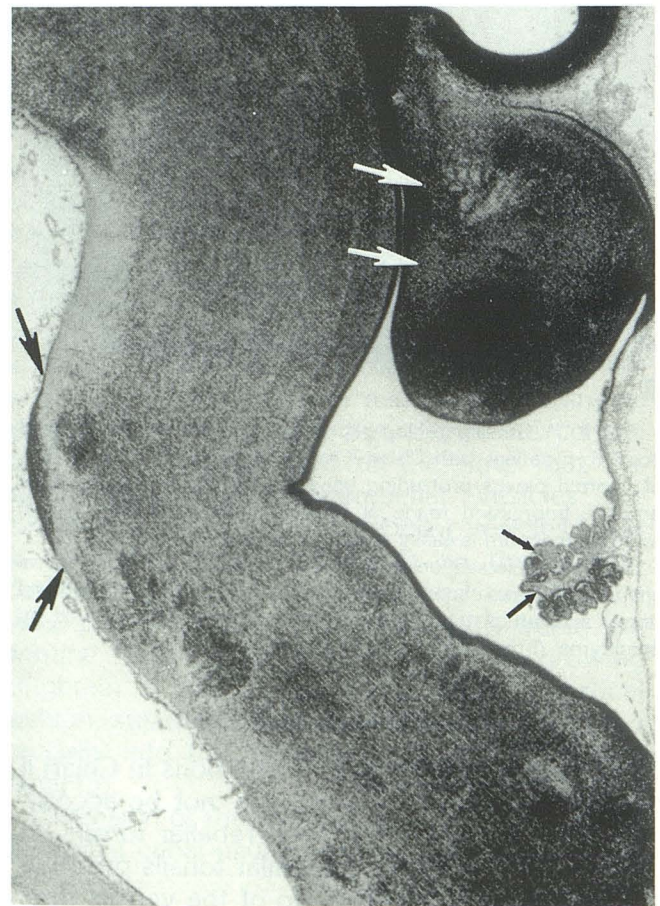


Fig. 4. Midline sagittal section through the hindbrain of a 45-mm human embryo. Cranial is to the left, and caudal to the right. Normal pontine flexure is present ventrally (*black arrows*). Note the intraventricular location of cerebellar tissue (*white arrows*) and the tuft of choroid plexus caudal to the cerebellum (*small black arrows*). (From Daniel and Strich (3).)

cerebellar peg in patients with Chiari II malformation should not be confused with a pathologic mass, but indeed is an expected finding in patients with spina bifida and the characteristic Chiari II hindbrain anomaly.

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