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*AJNR Am J Neuroradiol* 1991, 12 (4) 705-706

<http://www.ajnr.org/content/12/4/705.citation>

This information is current as of April 16, 2024.

## MR Imaging of an Extreme Case of Cerebellar Ectopia in a Patient with Chiari II Malformation

### Case Report

A 12-year-old boy who had been born with a meningomyelocele that was closed surgically 3 days after his birth had preoperative workup for a planned anterior release and posterior spinal instrumentation and fusion for worsening lumbosacral scoliosis. The patient had a history of developmental delay and learning disabilities and bowel and bladder incontinence. Physical examination showed thoracolumbar scoliosis with a curvature of 135° to the left. The lower extremities were spastic without evidence of any movements, deep-tendon reflexes were absent, and sensation was absent below the level of T6. MR of the spine and hindbrain showed cerebellar tonsillar herniation posteriorly and to the right of the cervical cord extending down to approximately the C3 level. From C3 to T3 on the left, an intradural extramedullary mass was present, which was not causing a mass effect on the atrophied spinal cord (Fig. 1A). This mass was dorsal and to the left of the atrophic lower cervical and thoracic cord (Fig. 1B) and did not enhance with gadopentetate dimeglumine. A CT myelogram confirmed this finding. MR of the brain showed absence of the entire left cerebellar hemisphere; the right cerebellar hemisphere was present in the posterior fossa, and the cerebellar tonsil was slightly ectopic (Fig. 1C). The patient had suboccipital craniectomy, cervical laminectomy, and intraspinal exploration to identify the cervical mass. At surgery, it was confirmed that the right cerebellar tonsil was ectopic as shown by MR; on the left side, attenuated vessels of vertebral artery origin coursed caudally, not in continuity with cerebellar tissue, but supplying the intradural extramedullary mass in the midcervical region. Biopsy of this mass showed that it was consistent with cerebellum. The vessels presumably arose from the posterior inferior cerebellar artery, and the mass represented a heterotopic cerebellar hemisphere.

### Discussion

The essential features of the Arnold-Chiari type II malformation include an elongated small cerebellum and brainstem, with caudal displacement of the medulla, parts of the cerebellum, and pons through an enlarged foramen magnum into the cervical spinal canal. Meningomyelocele and hydrocephalus usually are associated with

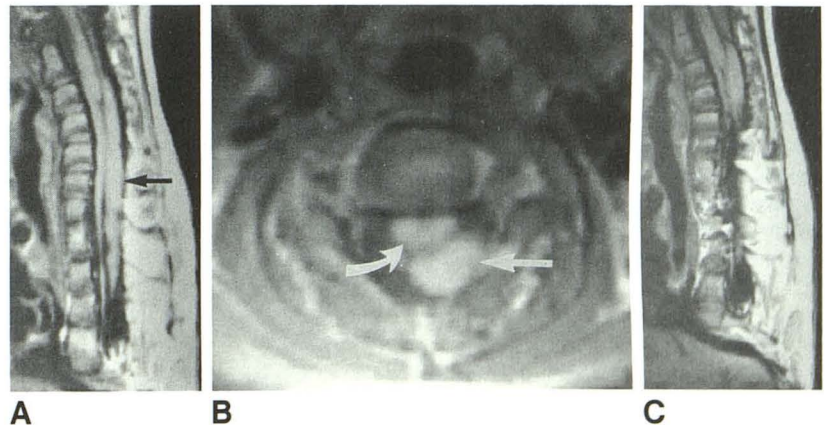
this anomaly. Other malformations of the neuraxis, including cerebral microgyria, cortical heterotopia, aqueductal stenosis, hydromyelia, syringomyelia, and diastematomyelia, have been reported [1].

Because of its superiority in depicting craniovertebral junction, MR is regarded as the best technique for showing structural abnormalities of the Chiari II malformation [2, 3]. Our patient's anomalies included an intradural extramedullary mass in relation to the dorsal cervical and thoracic cord, and biopsy showed that the mass was ectopic cerebellar tissue. This represented the absent left cerebellar hemisphere.

The presence of masses of nervous tissue in the cranial or spinal leptomeninges is a rare anomaly [4]. CNS heterotopias are neurons in abnormal locations and are thought to result from arrest of radial migration of the neuroblasts. They usually are confined to brain parenchyma. Heterotopias may be isolated anomalies or may be associated with other CNS malformations [5]. Chiari described congenital herniation of the brainstem and cerebellum in 1891. A gradation of severity of these malformations exists, ranging from severe infantile forms with gross skeletal abnormalities to adult forms in which the only abnormality is some degree of cerebellar tonsillar displacement [6]. El Gammal et al. [2] reported 19 cases of Chiari II malformation and one case of Chiari III malformation, all of which had caudal displacement of cerebellar tissue. In 10 patients, herniated vermis was clearly separate from the cerebellar tonsils. Curnes et al. [7] described 12 patients who had Chiari II malformation. In 10, the vermis was in the area of C2 to C4. In one patient, the vermis was at T1, and in another it was at T3. Wolpert et al. [3] described 24 patients with Chiari II malformations, including one in whom the underdeveloped cerebellum was almost completely in the upper cervical spinal canal. Mohr et al. [6] described 40 patients with primary cerebellar ectopia (adult Chiari malformation) who had a wide variety of clinical signs and symptoms. None of these reported cases had the severity of transforaminal tonsillar ectopia seen in our patient.

Our case had the typical features of a severe Chiari II malformation and an associated cerebellar heterotopia. Sagittal MR imaging of the craniovertebral junction resulted in visualization of the Chiari II malformation, the associated heterotopic cerebellar tissue, and the absence of the normal ipsilateral cerebellar hemisphere. This allowed preoperative speculation on the true identity of the mass. MR imaging

Fig. 1.—Cerebellar ectopia in Chiari II malformation.  
**A**, Contrast-enhanced sagittal MR image, 500/20/4, shows an intradural extramedullary mass (*arrow*) extending from C3 to T3. Surgery confirmed that mass was ectopic cerebellum.  
**B**, Contrast-enhanced axial MR image, 300/20/4, at level of C2 shows cervical cord (*curved arrow*) and ectopic cerebellar mass (*straight arrow*).  
**C**, Contrast-enhanced sagittal MR image, 800/20/4, shows contents of posterior fossa.



is an excellent method of demonstrating Chiari II malformations [2, 3, 7], particularly those of the severity described here.

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