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Spinal Intradural Cerebellar Ectopia

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Summary: An ectopic cerebellum, as in Chiari malformations and ectopic cerebellar dysplastic tissue, is a common finding; however, the presence of an organized ectopic cerebellum is exceedingly rare. We describe the MR imaging, surgical, and histologic appearance of an intraspinal ectopic cerebellum in an infant.

Intradural spinal cerebellar ectopias are commonly seen in the Chiari malformations (1). In the type I malformation, the cerebellar tonsils are located in the upper cervical spinal canal. In the type II malformation, the cerebellar vermis is herniated inferiorly, sometimes as far caudally as the thoracic spinal canal. These common ectopias are dorsal to the spinal cord and are always connected to the cerebellum (1). We present a case of an isolated cerebellar ectopia in the lower cervical spine. At MR imaging, an intradural extramedullary mass was seen, and at histologic examination, the mass showed cerebellar cortical cellular organization. We discuss the possible mechanisms responsible for this malformation.

Case Report

A 14-month-old boy presented with progressive scoliosis of 6 months' duration. On physical examination, his left foot was everted when he walked, his lower extremities were mildly hyperreflexic, and a positive right Babinski sign was elicited. Other developmental milestones were normal. Radiographs of the spine showed midline segmentation anomalies involving the T1-3 vertebrae and upper thoracic levoscoliosis (primary curve) with a secondary compensatory curve in the thoracolumbar region. MR imaging showed a mass in the spinal canal dorsal to the spinal cord at the C6-7 level (Fig 1A-D). The mass was isointense with spinal cord on both T1- and T2-weighted images and did not enhance after contrast administration. The spinal canal was widened, and inferior to the mass there was a cystic abnormality compressing the spinal cord from T1-8. The vertebral bodies of T-1 and T-2 showed central clefts and were slightly smaller in the anteroposterior dimension relative to the C-7 and T-3 vertebral bodies. The structures at the foramen magnum were normal. No abnormal cystic mass was seen anterior to the spine. The preoperative differential diagnosis included a hamartoma, dermoid, epidermoid, and an enteric duplication abnormality.

At surgery, a mass was seen within the dural sac, dorsal to the spinal cord (Fig 1E). On gross inspection, the surface of the mass resembled cerebellar hemispheres with densely packed

folia. Inferior to the mass, a cystic abnormality was adjacent to the compressed spinal cord. Its anterior wall was formed by the dorsal surface of the spinal cord and its lateral and posterior walls by thick arachnoid. The mass was resected in toto and the cyst decompressed and fenestrated to the spinal subarachnoid space. There was no obvious splitting of the spinal cord at the site of surgery. Light microscopy showed cerebellar cortex composed of outer and inner molecular granular cell layers (Fig 1F and G). Purkinje cells were present deep to these layers and an internal medullary layer was also present. Postoperative follow-up MR imaging showed no mass and adequate cyst decompression. Twelve months after surgery, the child's neurologic status remained unchanged.

Discussion

Apart from the common cerebellar ectopias associated with the Chiari I and II malformations, ectopic cerebellar tissue may be found in the fourth ventricle, or heterotopic cerebellar tissue may be seen in abnormal locations within the cerebellar parenchyma, but these ectopic or heterotopic tissues are usually microscopic and incidental in nature (2, 3). They are also found in the walls of occipitocervical encephaloceles and therefore may be associated with the Chiari III malformation (4). Other rare congenitally ectopic tissue found in the spinal canal has included renal tissue and a Wilms' tumor in association with a diastematomyelia (5, 6). Dermoids, epidermoids, and neurenteric abnormalities may also occur within the spinal canal (7).

The embryology of the cerebellar ectopia in our patient may be related to the so-called split-cord malformation, in which the developing notochord and neural tube are traversed by accessory alimentary canals (8). The tracts are composed of pluripotential cells and are able to differentiate into mesodermal tissue, resulting in diastematomyelia, enteric duplication anomalies, meningocele, lipoma, dermoid, epidermoid, and/or dermal sinus tracts (8). The spurs in diastematomyelia may be composed of bone, cartilage, fat, and/or fibrous tissue. These spurs result also in division of the thecal sac into two separate dural sheaths, leading to an external diastematomyelia. The endomesenchymal tract, however, may involute completely, leaving behind two hemicords within a single

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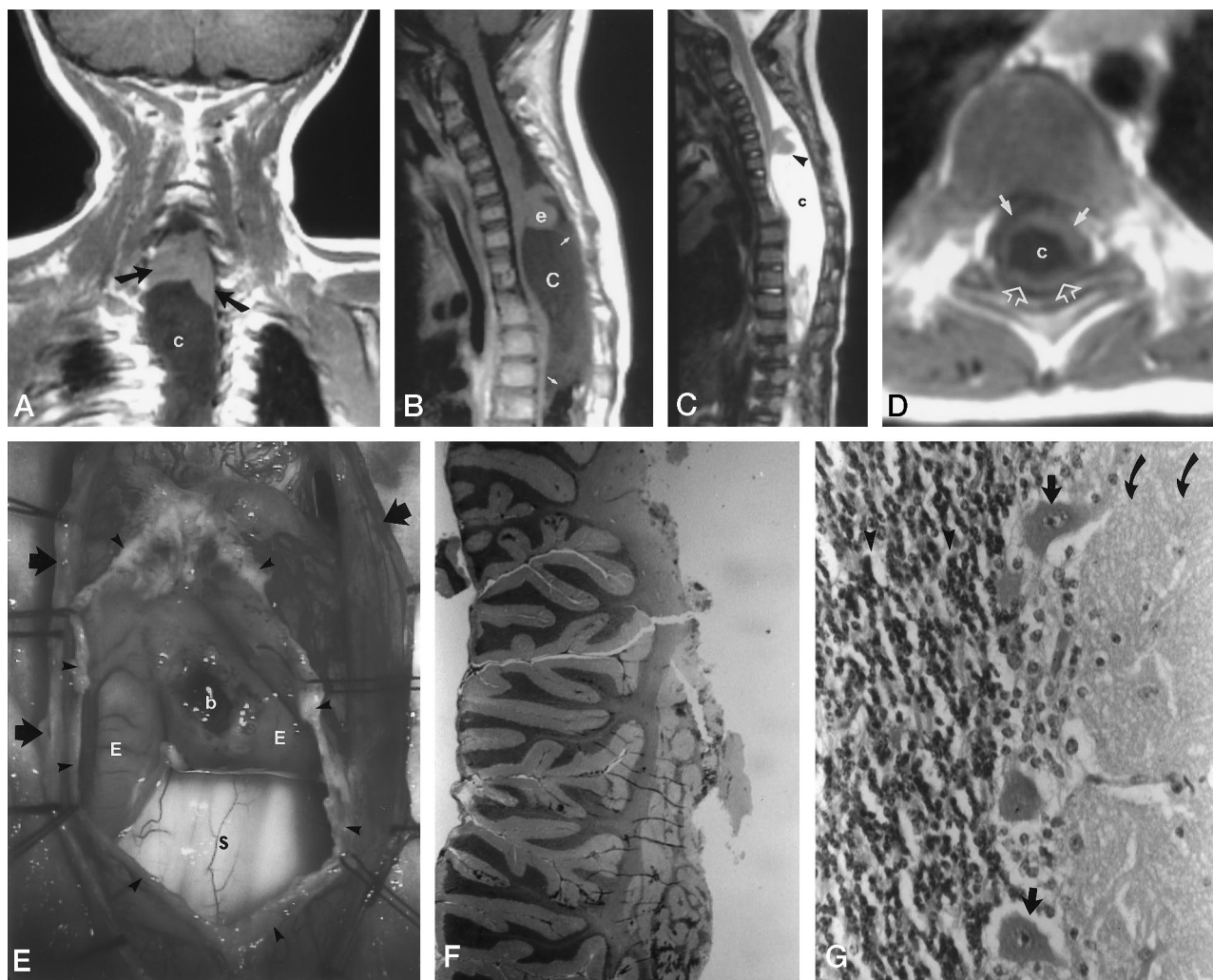


FIG 1. 14-month-old boy with heterotopic cerebellum.

A, Coronal T1-weighted MR image of the lower cervical and upper thoracic spine shows a bilobate mass (arrows) at the upper margin of the cystic cavity (c).

B, Midsagittal T1-weighted contrast-enhanced MR image shows absence of enhancement in the dorsal exophytic mass (e). The vertebral bodies of T-1 and T-2 appear slightly smaller in anteroposterior dimension relative to C-7 and T-3 vertebral bodies. The changes in these vertebral bodies are related to a central sagittal cleft of T-1 and T-2 vertebral bodies, which was evident on axial CT scans (not shown). C indicates cyst.

C, Corresponding sagittal T2-weighted MR image shows that the mass (arrowhead) is isointense with the spinal cord. The cyst (c) is bright, and inferiorly it contains some mixed signal intensity presumed to be related to pulsation artifact.

D, Axial contrast-enhanced T1-weighted MR image inferior to the mass shows the cyst (c) surrounded anteriorly by compressed spinal cord (solid arrows) and posteriorly by thick arachnoid (open arrows).

E, Intraoperative photograph of the heterotopic cerebellum. Paired ectopic cerebellar hemispheres (E) are seen in the dorsal aspect of the lower cervical spinal canal and correspond to the mass seen on MR images. The thick arachnoid (arrowheads) has been reflected and allows for visualization of the ectopia. Below the ectopia is the inner aspect of the cyst (its fluid has been removed). The dorsal surface (S) of the flattened spinal cord is well seen. The ectopia shows the biopsy site (b). The dura (arrows) is also reflected and tacked.

F, Low-power cross section of the resected mass shows typical cerebellar folia.

G, High-power image shows cerebellar cortical histology, including the granular cell layer (arrowheads), the Purkinje layer (straight arrows), and the molecular layer (curved arrows).

dural sheath and no spurs (an internal diastematomyelia) (8).

If communication between the primitive alimentary canal and the endomesenchymal tract persists, a neurenteric abnormality occurs (8). These abnormalities may include a simple diverticulum, a cyst, or a fistula, and may also be associated with intraspinal dermoids and lipomas (7, 8). Although these neurenteric anomalies generally involve the ventral spinal

canal, they may occur anywhere along the course of the endomesenchymal tract and occasionally may be located in the dorsal spinal canal posterior to the spinal cord. In addition, a persistent endomesenchymal tract may contain portions of the paramedian and median cell groups from the dorsal nerve roots. Thus, ganglion cells may become embedded in the dorsal region of a midline canal. These are believed to be responsible for the formation of the meningocele

manque, which is defined as a mixed bundle of nerve roots, fibrous bands, and blood vessels arising in an aberrant dorsal location of the spinal cord (8).

We believe that a similar mechanism may have accounted for the formation of the cerebellar ectopia found in our patient. That is, differentiation of pluripotential cells, including ganglion cells within the dorsal aspect of an endomesenchymal tract, led to the formation of a structure resembling the cerebellum. In all the above anomalies, including that in our patient, one may find midline vertebral body abnormalities (clefing), usually slightly inferior to the intraspinal anomaly (8). This slight difference in position may be explained by the differential growth between the spinal column and the cord. A cleft in the vertebrae is further evidence of a persistent canal originally between the amnion and yolk sac, traversing not only the neural tube but also the notochord, which is responsible for the formation of the mesenchymal elements of the spine. The fact that no true splitting of the spinal cord was found in our patient may be explained by complete involution of the endomesenchymal tract and the fact that the surgical exploration was limited to the site of the ectopic cerebellum. The caudal cystic cavity is difficult to explain. It was not a neurenteric cyst nor was it a syringomyelia, as only its ventral surface was formed by the spinal cord. Its lateral and posterior walls consisted of thick arachnoid. It is possible that the ectopic cerebellum resulted in obstruction to the normal flow of cerebrospinal fluid and to inflammation of the arachnoid membrane, resulting in a cyst.

Conclusion

We believe that this unusual case of intraspinal cerebellar ectopia may be part of the so-called split-cord malformation, as its presence and associated anomalies may be explained by a persistent endomesenchymal tract (containing ganglion elements) traversing both the notochord and neural tube early in life.

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