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MR of Lissencephaly

Lissencephaly is characterized by absent or poorly developed gyri, which are poorly seen on CT scans because of a bone artifact of the calvarium [1–4]. We report the use of MR imaging in a case of lissencephaly.

Case Report

MR imaging was performed on a 2-year-old girl whose history included low Apgar score, hypotonicity, and myoclonic seizures at birth. Her psychomotor and growth development were retarded. She had mild microcephaly and was hypotonic. Electroencephalography showed generalized epileptogenic activity.

MR imaging was performed on a 0.5-T scanner. T1-weighted images, SE 500/30, performed in the axial, sagittal, and coronal planes (Fig. 1) showed total absence of cortical sulci, apart from primitive sylvian, calcarine, and parietal occipital fissures.

Discussion

Lissencephaly is defined as smoothness of the brain's surface. It occurs in association with abnormalities in other parts of the body as part of a syndrome, such as the Miller-Dieker syndrome, or as an isolated abnormality [1, 5–8]. It may be inherited as an autorecessive chromosomal defect or may occur spontaneously without a genetic or familial history. Pathologically, the cortex may be completely agyric or may be polymicrogyric and pachygyric [1–4].

Microscopically, the gray matter only has four instead of the normal six layers. It has been postulated that the defect is the result of arrested migration at 80–90 days gestation [1, 4, 5]. As a result, the frontal and temporal opercula fail to develop, resulting in a widely open sylvian fissure and lateral ventricles that are dilated with a characteristic figure-of-eight configuration. The defective cellular migration often gives rise to heterotopic gray matter within the white matter at any location between the ventricles and the cortex [1, 5, 7]. The corpus callosum is often absent or hypoplastic. The brainstem also is involved and is hypoplastic [1, 5, 7, 8].

T1-weighted MR imaging is a sensitive method for evaluating the convexity and medial hemisphere gyri as well as sylvian fissures. The sagittal views appear to be the most useful in such topographic analysis. Microgyria cannot be diagnosed with certainty because of the limitations of resolution. Gray matter can be distinguished from white matter by using inversion-recovery and proton-density techniques, and it is theoretically possible to diagnose heterotopic gray matter, which sometimes is seen in this entity.

Calcification often is detected in white matter [1, 5]. The significance of this is unclear, but it may indicate viral infection as a cause of the lissencephaly in cases that have no clear-cut genetic cause [5]. MR is insensitive in imaging such calcification and should always be supplemented by CT scans.

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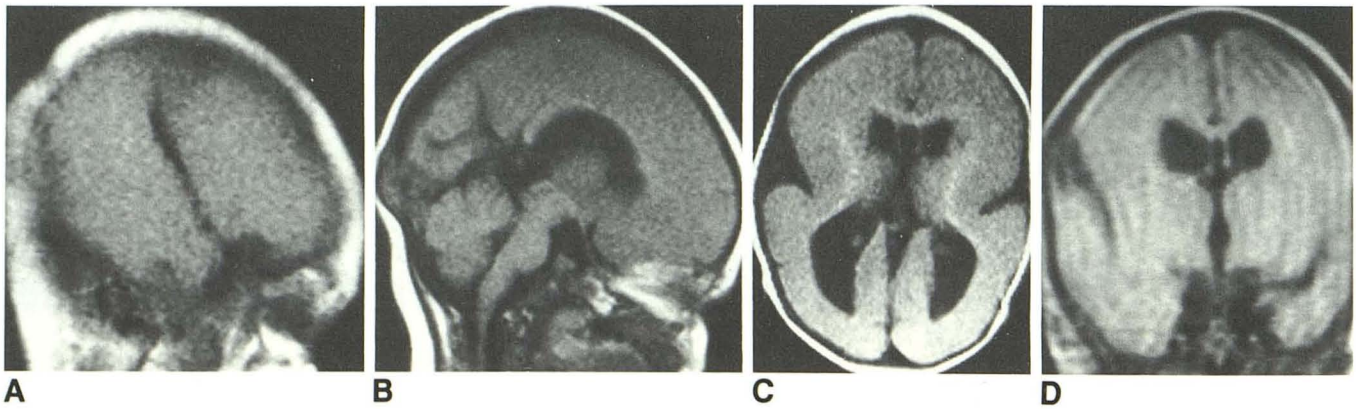

Fig. 1.—MR images, SE 500/30, of a 2-year-old girl with lissencephaly.
***A* and *B*, Sagittal images show total absence of gyri and sulci apart from sylvian fissure (*A*) and calcarine sulcus in the medial hemisphere (*B*).**
***C*, Axial image shows hypoplastic asymmetric sylvian fissures.**
***D*, Coronal image shows abnormal configuration of right sylvian fissure.**