Sonographic Recognition of Multiple Cystic Encephalomalacia

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Multiple cysts in both hemispheres were detected in four children under 1 year of age by real-time sonographic sector scanning. These cysts, typical of multiple cystic encephalomalacia, followed viral encephalitis in two, bacterial meningitis in one, and bacterial meningitis superimposed on intracerebral hemorrhage in one. The diagnosis of multiple cystic encephalomalacia, which has a grave prognosis, is readily made with high-resolution real-time sonography.

Multiple cystic encephalomalacia is a condition in which cerebral parenchyma is replaced by cysts of varying size. It has been described under a variety of synonyms in the pediatric and pathologic literature [1–11]. Occasionally present at birth [1], it is seen in the first year of life and is associated with profound neuromotor delay. The cystic deformity has been recognized in life by ventriculography [2], pneumoencephalography [3], and computed tomography (CT) [4, 5].

The condition can be recognized readily by cranial sonography while the anterior fontanelle is open using a 5 MHz sector scanner. We have seen four infants with CT correlation in two and autopsy correlation in one. The prognosis is poor, and prompt recognition of the disease will facilitate timely family counseling.

Case Reports

Case 1

A 9-day-old boy delivered at term was admitted to Arkansas Children's Hospital because of poor feeding, irritability, and fever. Cerebrospinal fluid (CSF) culture grew *Enterobacter cloacae*.

Despite treatment with appropriate antibiotics, fever persisted and the infant became lethargic and apneic and required ventilator therapy. Cranial sonography (Advanced Technology Labs., Bellevue, WA) showed periventricular cavitation and hydrocephalus (fig. 1A). Ventricular enlargement was successfully treated by shunting. The intracerebral cysts enlarged over the next 6 months so that only strands of cerebral tissue remained (fig. 1B). The infant remained severely retarded with little subsequent change in cranial sonograms.

Case 2

A male infant delivered at 32 weeks gestation was transferred to Arkansas Children's Hospital at 4 days of age after severe postnatal acidosis and asphyxia. Cranial sonography showed extensive bilateral intracerebral bleeding (fig. 2A). The CSF contained protein of 258 mg/dl; glucose, 1 mg/dl; and a white blood cell count of 2,500/mm^3, 90% of which were neutrophils. CSF culture grew *Bacillus cereus*. Despite appropriate antibiotics, the child became profoundly retarded. Repeated cranial sonographic studies showed multifocal cavitation in both hemispheres that became more extensive in the course of the following months (fig. 2B).

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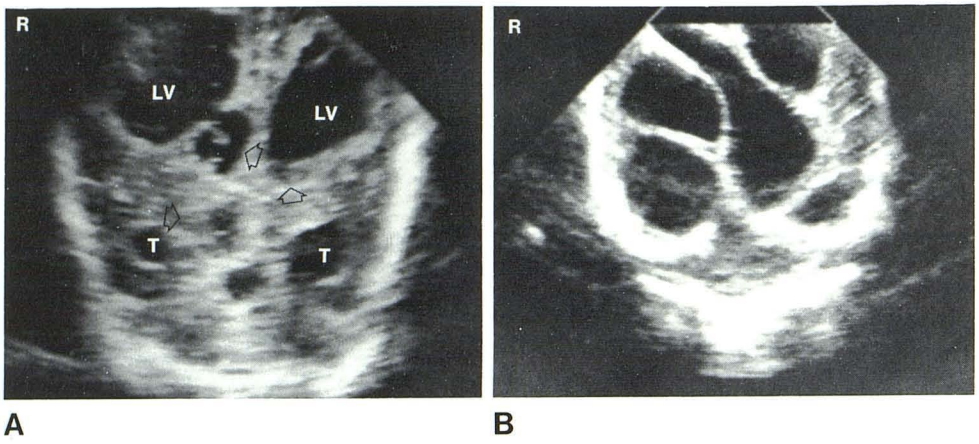

Fig. 1.—Case 1. Coronal sonograms. **A**, Dilated irregular lateral ventricles with smaller cavities (*arrows*) in surrounding brain. LV = body of lateral ventricle; T = temporal horn of lateral ventricle. **B**, 6 months later. Almost complete replacement of brain by cysts. Remaining strands of brain undulate loosely in cyst fluid. Ventricles no longer discernible.

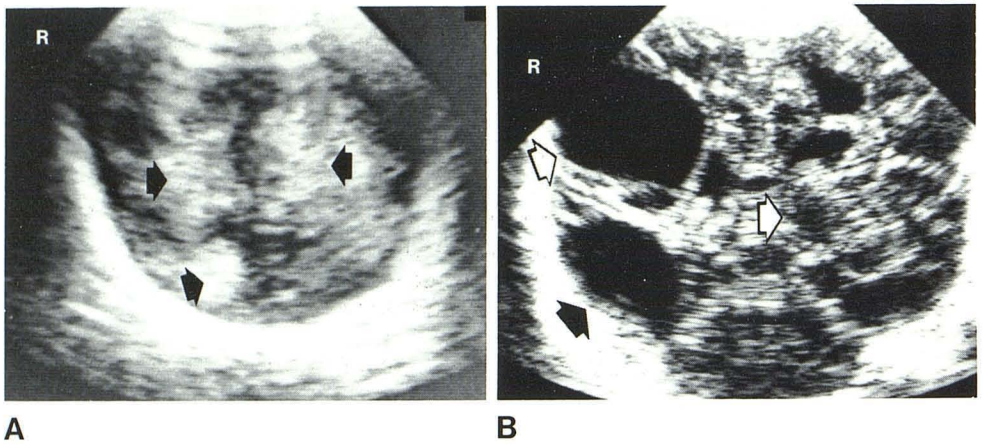

Fig. 2.—Case 2. Coronal sonograms. **A**, Severe bilateral intracerebral hemorrage (*arrows*). **B**, 6 months later. Irregular asymmetric cavitation (*arrows*) typical of multiple cystic encephalomalacia. Ventricular system is compressed and difficult to see.

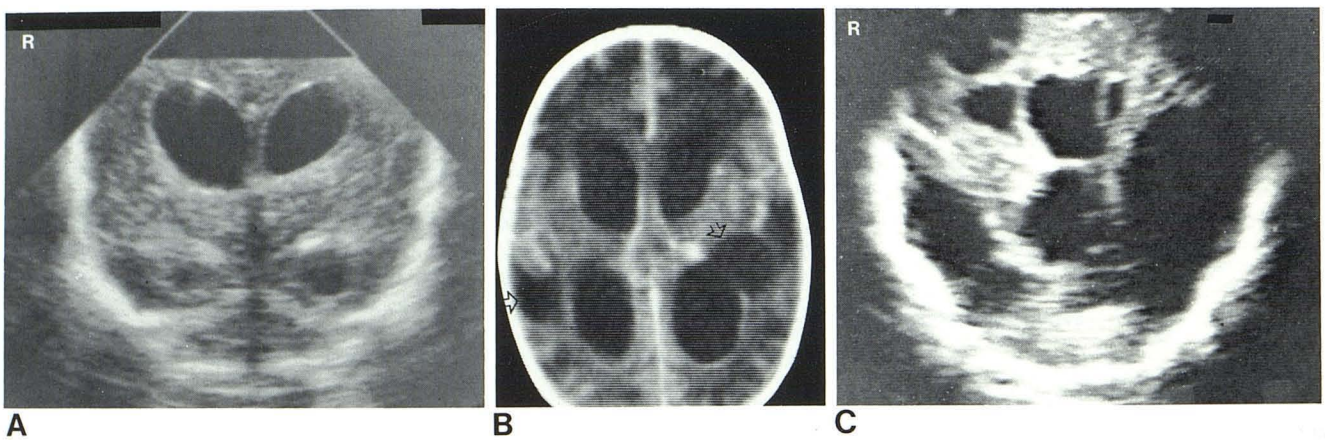

Fig. 3.—Case 3. **A**, Coronal sonogram. Moderate symmetric hydrocephalus. **B**, Axial CT scan. Ventricular dilatation and widespread periventricular low-density areas suggestive of encephalomalacia. Frank cysts lie lateral to atria (*arrows*). **C**, Coronal sonogram 3 months later. Asymmetric irregular cavitation in both hemispheres. Ventricles are no longer dilated but are compressed by cysts and not well shown.

Case 3

A 2-month-old boy presented with failure to thrive and an enlarged head. Clinical assessment revealed a positive Moro reflex, inability to hold the head, and a very poor suck reflux. Cranial sonography showed hydrocephalus (fig. 3A). CT also showed large ventricles and periventricular areas of low attenuation (fig. 3B). The serum antibodies for herpes simplex 2 were significantly increased.

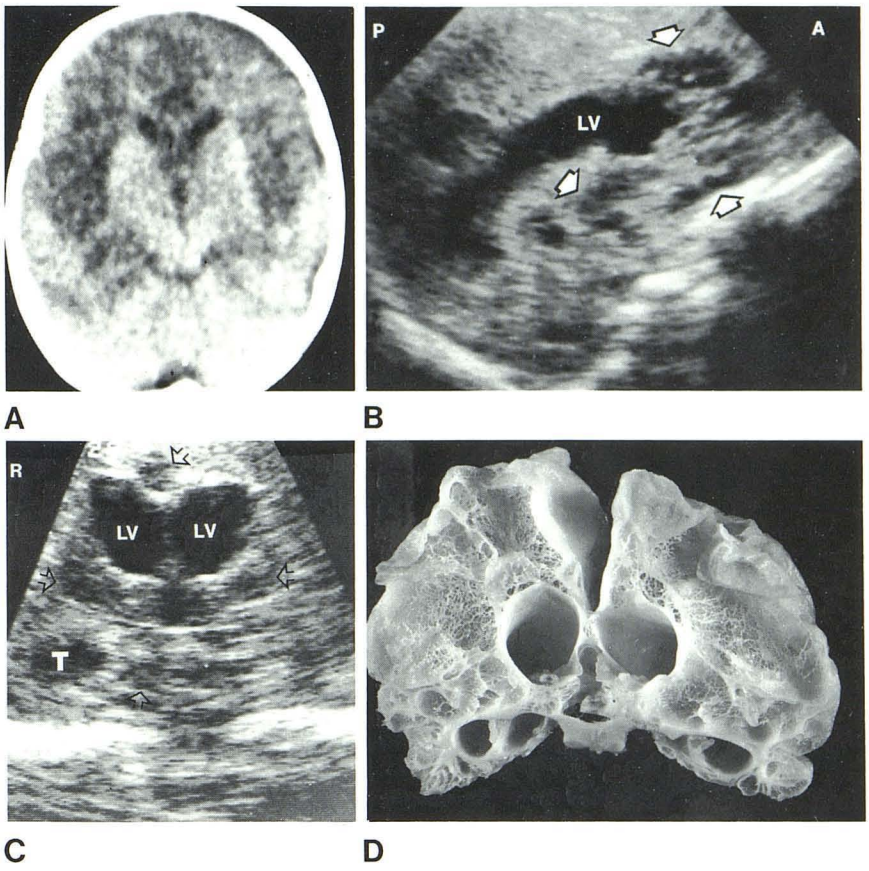

Fig. 4.—Case 4. **A**, Axial CT scan. Periventricular low-density areas suggestive of encephalomalacia. **B**, Sagittal sonogram. Irregular periventricular cavities (*arrows*) suggestive of multiple cystic encephalomalacia. A = anterior; P = posterior; LV = body of right lateral ventricle. **C**, Coronal sonogram. Moderate ventriculomegaly and numerous poorly defined anechoic areas in periventricular parenchyma and in basal ganglia (*arrows*). T = temporal horn of lateral ventricle. **D**, Coronal brain section. Parenchyma of both hemispheres replaced by honeycomb of fine cavities.

Subsequent sonographic examinations showed resolution of the hydrocephalus and the formation of numerous irregular cysts. A particularly large cyst on the right was drained with a cystoperitoneal shunt. A cyst on the left continued to enlarge (fig. 3C). Later the shunt became blocked and the porencephalic cysts on both sides enlarged further. After failing to appear for further cystoperitoneal shunts, the child died at home. There was no autopsy.

Case 4

A 7-month-old girl presented with a 24-hr history of seizures. CT showed irregular periventricular areas of low density (fig. 4A), and a diagnosis of encephalitis was entertained. CSF culture grew cytomegalovirus.

High doses of anticonvulsants were required for persistent seizures. She developed marked neuromotor retardation. Cranial sonography showed numerous irregular anechoic cavities about the third ventricle (figs. 4B and 4C). Follow-up examinations over the next 6 months showed little change. The child remained profoundly retarded and died of an aspiration pneumonia at age 16 months.

Autopsy revealed innumerable cysts throughout both hemispheres (fig. 4D) but with sparing of the brainstem and cerebellum. Histopathologic examination revealed severe demyelinization, reactive gliosis, and chronic inflammation with foci of granulomatous reaction and dystrophic calcification. Large mononuclear cells and giant cells with intranuclear inclusion bodies suggestive of cytomegalovirus were seen.

Discussion

Multiple cystic encephalomalacia is an important disease to recognize because the prognosis is so poor. Further, in the neonate, it may be clinically silent; only when the infant fails to achieve his early milestones of neuromotor development may the severe functional deficiency become apparent.

Multiple cystic encephalomalacia may follow a variety of insults to the central nervous system including asphyxia [2, 5], meningitis [4], encephalitis [7, 8], and twin-to-twin transfusion [1, 3, 9]. Our four cases diagnosed by sonography seem to be examples of the first three. The common factor linking these disparate conditions appears to be brain hypoxia [10, 11].

Cranial sonography should be considered in the assessment of all neonates and infants who suffer from asphyxia, intracerebral hemorrhage, and intracranial infection. As the prognosis of multiple cystic encephalomalacia is grave, prompt recognition with sonography may affect further management and family counseling. Enlargement of the cysts can be detected and the effectiveness of cyst taps or cystoperitoneal shunts recorded. Our experience suggests that the cysts often do not communicate. To prevent inordinate increase in head size, more than one cyst shunt may be required.

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