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Schizencephaly—Imaging by MRI

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Schizencephaly is a congenital brain anomaly of neuronal migration characterized by holohemispheric grey matter lined clefts. As it forms a migrational disorder there may be associated anomalies. MRI due to its superior soft tissue contrast resolution and multiplanar imaging capability has been reported to be an ideal modality for total evaluation of this anomaly(1-5). We here describe our MRI findings in

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three infants of this relatively uncommon condition.

Case Reports

A retrospective analysis of three infants of schizencephaly imaged on MRI was made. All the infants were studied by 1.5 Tesla superconducting system in head surface coil in supine position. Patients were sedated by syrup triclofos. A slice thickness of 5 mm and interslice gap of zero (for sagittal) or 50% (for axial) was used. The study was performed in spin echo sequence using TE/TR of 22/500-700 msec for T1 and TE/TR of 90/2000 msec for T2 images. Images were acquired on 256 × 256 matrix.

Case 1:

A 47-day-old female was referred with primary complaint of recurrent seizures. MRI revealed partial agenesis of right parietal lobe with complete clefting. The cleft was extending up to the lateral ventricle. There was a large cyst in the posterior fossa communicating with the fourth ventricle with almost complete agenesis of cerebellum (Dandy Walker cyst). There was associated partial agenesis of corpus callosum (*Fig. 1*).

Case 2:

A 4-year-old male presented with recurrent seizures and delayed milestones. MRI revealed bilateral schizencephaly with complete clefting on right and incomplete clefting on left side. Images also revealed heterotopia in the left parietal lobe giving same signal intensity as cortex in T1 as well as on T2 images. This was seen just medial to the site of clefting. There was evidence of polygyria also on right side. (*Fig. 2*).

Case 3:

MRI in a 2-year-old female child pre-



Fig. 1. Axial T1 weighted image showing right sided schizencephaly and associated Dandy Walker malformation.



Fig. 2. Axial T2 weighted image showing bilateral schizencephaly with heterotopia in the left parietal lobe.

senting with delayed milestones and chronic seizures revealed right schizencephaly with complete clefting. No other associated anomaly was detected.

Discussion

Neuronal migration is a highly complex and organized step in brain development. The process is lengthy and complex inviting major vulnerability to a host of possible insults. Disturbances of neuronal migration are usually characterized by abnormal development of the cortical mantle, so that it is too thick, too flat or too folded.

Schizencephaly refers to full thickness clefts within the cerebral hemispheres. These clefts pathologically, are characterized by an infolding of grey matter along the cleft from the cortex into the ventricles and there may be fusion of the cortical pia and ventricular within cleft, the so called pia-ependymal seam(6).

MRI was helpful in demonstrating the cleft going upto ventricles and in showing associated anomalies—Dandy Walker syndrome, heterotopia, polygyria and agenesis of corpus callosum. The various anomalies associated with schizencephaly reported earlier are—absence of septum pallidum, dysgenesis of corpus callosum, polymicrogyria and ependymal heterotopia(2,5,6).

The exact etiopathogenesis of schizencephaly is not firmly established. It is believed to be caused by complete agenesis of a section of cerebral tissue. Barkovich and Norman(2) hypothesized it is due to an episode of hypotension, causing infarction of the watershed area. They said areas of polymicrogyria and heterotopia commonly seen surrounding the cleft are secondary to ischemic changes in the less severely affected areas. In our Case 2 also, the heterotopia was seen close to the cleft.

On imaging modalities it is essential to differentiate between schizencephaly and porencephaly. Knaap and Volk(7) reserved the term schizencephaly and porencephaly

for clefts in the brain with or without hydrocephalus, with or without fused lips, either uni or bilateral and either developmental or encephaloclastic. Porencephaly is reserved for a cavity within the cerebral hemisphere that does not interconnect the lateral ventricle and the subarachnoid space.

We feel from these cases that MR provides excellent demonstration of the anatomic changes in schizencephaly and MR should be the imaging method of choice in the evaluation of these anomalies.

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NOTES AND NEWS

WORKSHOP ON GROWTH MONITORING AND NUTRITION SURVEILLANCE

A Workshop on 'Growth Monitoring and Nutrition Surveillance Problem' will be organized by the Growth and Development Chapter and Nutrition Chapter of IAP Sub-speciality on *17th January, 1993* during the XXX National Conference of IAP, Calcutta. The Registration Fee is Rs. 100/- which is to be paid by Demand Draft drawn on 'XXX National Conference of IAP, Calcutta' payable at Calcutta and to be sent to the Organizing Secretary at 9/1 Ramanath Pal Road, Calcutta 700 023.

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