Childhood convulsion and hemiparesis due to schizencephaly-a case report

M Salek, H Alam, M Islam, M Zaman

Citation

Abstract
Childhood convulsion with hemiparesis due to schizencephaly is a rare condition. It is a developmental disorder of neuronal migration, characterized by early focal destruction of the germinal matrix and surrounding brain before the cerebral hemispheres are fully formed at 1-5 months of gestation. The lesion is most likely related to multiple aetiologies including genetic, toxic, metabolic, vascular or infectious agents. This case is reported due to its worldwide rarity.

INTRODUCTION
Schizencephaly, a developmental disorder of neuronal migration is characterized by cleft in cerebral mantle, which communicates between the subarachnoid space laterally and ventricular system medially. This disorder was originally described by Wadsworth and Yakolev. Their original work describes schizencephaly to result from failure of normal migration of primitive normoblasts resulting in cerebral cleft. The cardinal neuropathological features are i) Hemispheric cleft ii) communication of subarachnoid space with lateral ventricle medially iii) Infolding of grey matter along the cleft iv) Multiple associated intracranial malformations including polymicrogyria, absent septum pellucidum, optic nerve hypoplasia. The presentation and outcome are variable i.e. hemiparesis, developmental delay, microcephaly, mental retardation and most patients have seizures.

CASE REPORT
A 2 year aged female child was admitted in paediatric neurosurgery ward of CM Hospital Dhaka with complain of repeated episode of convulsion since birth and weakness in right upper and lower limbs since two months of age. She was symptomatically treated with anticonvulsant sodium valproate.

The child was delivered full term normally at home with unremarkable antenatal maternal history. She cried soon after birth and there were no postnatal complications. There is no history of consanguineous marriage. On physical examination child had normocephaly, no dysmorphism of face. Anterior fontanelle was normal. Sutures were widely normal. There was a global delay in the developmental milestones. Muscle tone of the right upper and lower limbs showed spasticity with 4/5 power grade with normal reflexes. Vitals were normal. Anthropometric measurements of weight and length were within normal limits. The child was admitted with provisional diagnosis of Cerebral palsy. Investigations done revealed normal Hb, Tc, Dc. EEG showed abnormal record with seizure pattern. CT scan showed SCHIZENCEPHALY.

Figure 1
Fig: CT scan of brain to show schizencephaly

Thus the diagnosis was established with the help of CT scan report. The option of treatment with prognosis was explained to the parents. She eventually underwent cystoperitoneal shunt. Postoperative recovery was uneventful except an episode of seizure that was managed by sodium valproate.
Counseling was given and physiotherapy was advised. Anticonvulsant was continued.

DISCUSSION

Schizencephaly is a rare developmental disorder of neuronal migration. It is characterized by CSF filled cleft, lined by grey matter and extends across the entire cerebral hemisphere from ependyma to the periphery of the brain. It is a rare developmental disorder of neuronal migration. It is characterized by CSF filled cleft, lined by grey matter and extends across the entire cerebral hemisphere from ependyma to the periphery of the brain. Types of Schizencephaly:

- Type I: Closed type (lips of cleft are fused)
- Type II: Open type (clefts are separated)
- Type II is more common than type I

INCIDENCE

It is present at birth with no sex predilection. The lesion is related to multiple aetiologies including genetic, toxic, metabolic, vascular or infectious agents such as cytomegalovirus infections. As genetic cause, it is found to have a mutant gene, EMX2.

CLINICAL FEATURES

The symptoms of schizencephaly are variable. Severity is related to the extent of cortex involved in the defect:

- Mild hemisparesis and seizure with normal development.
- Mild to moderate developmental delay with hemiparesis.
- Severe mental deficits, severe motor anomalies including spastic quadraparesis. Frequently these patients present with blindness often associated with optic nerve hypoplasia.

INVESTIGATIONS

MRI is the imaging modality of choice due to its superior differentiation of gray matter and its ability to image in more than one plane. Identification of gray matter lining the cleft is the pathognomonic finding in differentiating schizencephaly from disorders like porencephaly. Other modalities of choice are CT Scan, Ultrasound but the degree of confidence is not as high as MRI.

TREATMENT

It consists of treatment of seizures, physiotherapy, and in cases that are complicated by hydrocephalus, a ventriculoperitoneal shunt is needed.

PROGNOSIS

It depends upon the amount of brain involved. In open lip schizencephaly the patients die at early age. Death is mainly due to failure to thrive, chronic infections and respiratory problems. Whereas in closed lip schizencephaly patients may not present until later in infancy and they live up to adulthood.

CONCLUSION

Schizencephaly is a rare developmental disorder of cerebral cortical development. It mainly manifests as two types - open type and closed type. Most of the cases (80-90%) present with absent septum pellucidum. Presentation is variable with poor prognosis with symptomatic treatment.

References

4. Capra-V; De-Marco-P; Moroni-A; Faiella- A; Brunelli-S; Tortori-Donati-P; Andreussi- I; Boncinelli-E; Cama-A Schizencephaly: Surgical features and new molecular genetic results. Emr J Pediatr Surg 1996 Dec; suppl 1:27-9
Author Information

MAA Salek, FCPS, MRCS
Department of Neurosurgery, CM Hospital

HMS Alam, FCPS, FICS
Department of Neurosurgery, CM Hospital

MA Islam, FCPS, MS
Department of Neurosurgery, CM Hospital

MR Zaman, FCPS, MS
Department of Neurosurgery, CM Hospital