Schizencephaly In Children: Clinical Features And Its Complications

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Background

Schizencephaly is a rare congenital central nervous system malformation. Clinical features of schizencephaly vary widely ranging from developmental delay, microcephaly, hydrocephalus, spasticity, epilepsy, endocrine abnormalities in the HPA axis and visual abnormality.

Objectives

We aim to describe clinical features and complication of schizencephaly in children at Siriraj hospital, a major tertiary referral center in Thailand.

Methods

A retrospective chart review was performed on all patients, aged less than 15 years, diagnosed with schizencephaly in pediatric neurology clinic at Siriraj hospital during January 2005 to December 2015. We identified from a hospital database by using the ICD-10.

Results

There were 75 patients included in the study. Bilateral open-lips schizencephaly is the most common type and found in 35 patients (46.7%) and followed by unilateral open-lips (21 patients, 28%) and unilateral closed-lips schizencephaly (10 patients, 13.3%). The most common clinical features of schizencephaly were motor disability (92%), other developmental disability (86.7%), and epilepsy (45.3%). Associated CNS findings are absent of septum pellucidum in 60/75 (80%), optic nerve hypoplasia in 32/63 (50.7%) and pituitary hormone abnormalities in 12/49 (24.5%). Septo-optic dysplasia was diagnosed in 33 patients (44%).

Conclusion

Bilateral open-lips schizencephaly has the worst overall outcome in terms of motor deficits, other developmental disability and epilepsy. We recommend that if schizencephaly is found on neuroimaging study, other CNS abnormalities such as ASP, polymicrogyria, and dysgenesis of corpus callosum should carefully be looked for. A formal ophthalmologic examination and pituitary hormone abnormalities should be considered since SOD may be found in up to 44% among patients with schizencephaly.

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