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Original Research Paper

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HEMIPLEGIA HEMICONVUSION HEMIATROPHY SYNDROME-A CASE REPORT

Dr Anuj Nehete	Post graduate, Department of Neurology JSS Hospital, Mysuru.
Dr Shasthara. P*	Post graduate, Department of Neurology JSS Hospital, Mysuru. *Corresponding Author
Dr Harsha S.	Professor and Head, Department of Neurology JSS Hospital, Mysuru.

ABSTRACT Neuronal migration disorder is a rare cause of seizure, weakness and developmental delay[1]. Individuals with smaller, unilateral clefts may be paralyzed on one side of the body[2]. Outcome and presentation of schizencephaly are variable, but it typically presents with seizures ,hemiparesis,and developmental delay. Here we report a case of open lip schizencephaly presenting as Hemiplegia Hemiconvusion Hemiatrophy Syndrome.

KEYWORDS : schizencephaly,Neuronal migration disorder.

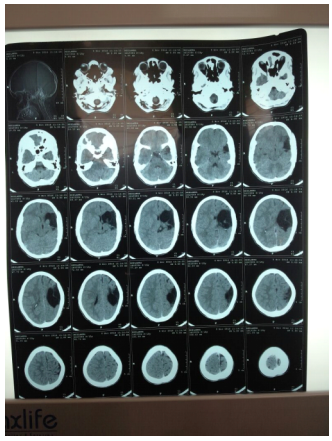
INTRODUCTION

Seizures, Hemiplegia,Hemiatrophy irrespective of etiology, has been considered As HHE syndrome[3].Prolonged ictal activity that act on blood-brain-barrier permeability, genetic factors and focal epileptogenic lesion predispose to this syndrome. In type I or closed-lip schizencephaly, the cleft walls are in apposition and type II or open lip schizencephaly, in which the walls are separated. Schizencephaly type II occurs more commonly than type I.

CASE REPORT

14 yr old female born of non-consanguineous marriage presented with the history of infantile hemiplegic,recurrent seizures involving right half of body, and wasting of right half of body with mental retardation. Her antenatal-perinatal-family history was uneventful. In view of diagnosis of Hemiparesis-Hemiatrophy-Hemiconvulsion syndrome,she was subjected to CT brain which showed Open lip type of schizencephaly. She was managed with physiotherapy, two anticonvulsants and is doing fine at follow up.

Clinical photograph of patient showing right infantile hemiplegia and hemiatrophy.



CT brain of patient showing open lip schizencephaly on left side.

DISCUSSION

Schizencephaly is an extremely rare congenital brain anomaly and is the most severe form of neuronal migration defect4. schizencephalic clefts denote defects occur early in the second to fifth month gestation, prior to the end of neuronal migration. There is very scant literature on schizencephaly in Indian population.

CONCLUSION

Motor deficits are the predominant manifestations in open-lip schizencephaly.In patients of congenital hemiparesis,hemiatrophy, hemiconvulsion one should consider the presence of neuronal migration disorders like schizencephaly.

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