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SCHIZENCEPHALY: A CASE REPORT

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ABSTRACT

Schizencephaly, sometimes called agenetic porencephaly, is the term used to describe gray-matter lined clefts that extend through the entire cerebral mantle, from the ependymal lining of the lateral ventricles to the pial covering of the cortex. A 1 year old male child was admitted with the history of delayed developmental milestone and spasticity of both upper and lower limbs since birth. The proper antenatal and natal history could not be obtained as the child was adopted. He was previously diagnosed as a case of cerebral palsy. On physical examination, the child had microcephaly, no dysmorphism of face. Anterior fontanelle was normal. Sutures were normal and there was a global delay in the developmental milestones. Muscle tone of the upper and lower limbs showed spasticity with normal reflexes. Vitals signs were normal. Anthropometric measurements of weight and length were within normal limits. Plain CT scan of the brain showed wide CSF filled cleft in left frontal lobe and another narrow cleft in right parieto temporal lobe extending from cortex to lateral ventricles. The child was diagnosed as a case of bilateral schizencephaly. The incidence of schizencepaly is very low 1.5 in 1,000,000 live births. Only few cases have been described in the literature. The etiology of such phenomenon is also poorly understood.

KEYWORDS : Schizencephaly, Cerebrospinal fluid, Microcephaly, Dysmorphism, Anterior fontanelle, Anthropometric measurement, CT scan

INTRODUCTION:-

Schizencephaly, sometimes called agenetic porencephaly, is the term used to describe gray-matter lined clefts that extend through the entire cerebral mantle, from the ependymal lining of the lateral ventricles to the pial covering of the cortex.[1] It is a neuronal migration anomaly, caused by insults to migrating neuroblasts during 3rd to 5th gestational months.

This disorder was originally described by Wadsworth and Yakolev.[2] Their original work describes schizencephaly to result from failure of normal migration of primitive normoblasts resulting in cerebral cleft. Two types are recognized, which have prognostic significance. 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.[3] In either instance the cleft is lined by heterotopic gray matter. Although no specific cause has been identified, various hypotheses have been postulated. It is thought to be due to abnormal neuronal migration or localized ischemia.[4] Other causes are the expression of genetic factors such as the mutant gene, EMX2. These expressed genetic factors are believed to damage the periventricular germinal matrix impairing cellular migration at 6–7 weeks of intrauterine growth. Sarnat and Curatolo, in his research, described it as an extreme form of true porencephaly due to the ischemic theoryMagnetic resonance imaging (MRI)[5] is the imaging modality of choice for its diagnosis in infants and children due to its high tissue resolution and nonionizing attribute. A study done by Liang et al.[6] found a correlation between imaging. findings and clinical features of schizencephaly thus helping to predict the developmental outcome. MRI allows for the visualization of the gray matter which lines the clefts which is pathognomonic for this condition differentiating it from porencephaly as well as in identifying other acquired and associated lesions. The incidence of schizencepaly is very low 1.5 in 1,000,000 live births. Only few cases have been described in the literature. The etiology of such phenomenon is also poorly understood.[7]

Figure1

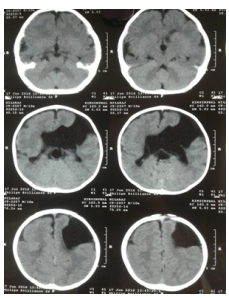
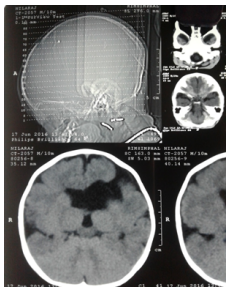


Figure 2



CASE REPORT:-

A 1 year old male child was admitted with the history of delayed developmental milestone and spasticity of both upper and lower limbs since birth.The proper antenatal and natal history could not be obtained as the child was adopted .The child had microcephaly but no facial dysmorphism .Sutures were normal and there was a global delay in the developmental milestones.Muscle tone of the upper and lower limbs showed spasticity with exaggerated reflexes.Hip abduction was restricted to 20 degree bilaterally with popliteal angle of 55 degree on both side.Anthropometric measurements of weight and length were within normal limits . A working provisional diagnosis of cerebral palsy was made.Laboratory investigations: Packed cell volume, electrolytes, thyroid profile all were within normal limit. Plain CT scan of the brain showed wide CSF filled cleft in left frontal lobe and another narrow cleft in right parieto temporal lobe extending from cortex to lateral ventricles . The child was diagnosed as a case of bilateral schizencephaly. Counseling of the parents about the disease and prognosis was done. The patient was managed conservatively. Neurodevelopmental training including rolling technique for prone lying and balance board was given.

DISCUSSION :-

Schizencephaly is an extremely rare congenital brain anomaly and it is the most severe form of neuronal migration defects. Etiologies include intrauterine infections, maternal trauma of several types, teratogens, alcohol and drug abuse, warfarin, and monozygotic twin interactions.Clinical presentation depends on the size and location of the lesion. Prognosis for individuals with schizencephaly varies depending on the size of the clefts and the neurological deficit degree Generally treatment for individuals with schizencephaly usually consists of physical therapy, treatment for seizures. In a few cases if there was complication of hydrocephalus, a shunt is required

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