INTRODUCTION:

Hemiparesis occurs due to various etiologies such as infarction, malformations, cortical dysplasia and gliosis. Schizencephaly is extremely rare with an estimated incidence of 1.5 per 100,000 live births which may present as hemiparesis. Schizencephaly is a congenital disorder of cell migration with defect in sulcation. It is characterized by cleft in cerebral mantle, which communicates between ventricular system medially and subarachnoid spaces laterally. In type I or closed-lip schizencephaly, the cleft walls are in apposition and type II or open lip schizencephaly, the cleft walls are separated. Motor deficits are the predominant manifestations in open-lip schizencephaly. Neuroimaging is useful in locating the defect and associated malformations.

CASE HISTORY:

7 year-old girl born for third degree consanguineous parents presented with weakness of left upper and lower limb since birth. There was no H/O of seizures or trauma. Child was born at term, by vaginal delivery. Antenatal, natal, and postnatal periods were uneventful. Child had global developmental delay. On examination child had left hemiparesis with mild deviation of angle of mouth towards left and left upper limb atrophy (Figure 1). Ophthalmic and ENT evaluation was normal. Patient had subnormal intelligence. The rest of the systemic examination was normal. CT of brain (Figure 2) showed CSF filled cystic lesion noted in right precentral gyrus lined by grey matter and communicating with right lateral ventricle, absent septum pellucidum features consistent with open lip schizencephaly.
DISCUSSION:

Schizencephaly is the most severe form of neuronal migration disorder. The clefts can be unilateral or bilateral, symmetric or asymmetric. Only one-half of the schizencephaly cases are bilateral. In bilateral, only 20% are of mixed type (type I and II). Type II occurs more common than type I. They can appear anywhere in the brain, although they are usually perisylvian. In either instance the cleft is lined by gray matter. Presentation and outcome of children are related to extent of cortex involved. The presence of schizencephalic clefts lined by grey matter suggests that these defects occur early in the second to fifth month gestation, prior to the completion of neuronal migration. Etiologies include in utero infections - cytomegalovirus and herpes virus. Other etiologies include teratogens, alcohol and drug abuse, warfarin, and monozygotic twin interactions. In our case, there were no stigmata of congenital infections. Clinical presentation depends on size and location of the lesion. Bilateral clefts are generally associated with quadriplegia and severe cognitive impairment. MRI identifies the anomalous grey matter along the cleft as well as the associated abnormalities.

CONCLUSION:

This case is reported for its rarity. Congenital hemiparesis one should consider the presence of neuronal migration disorders like schizencephaly. Appropriate diagnosis is necessary for counselling and for offering prognostic information to parents.

REFERENCES: