

AN INTERESTING CASE REPORT OF PORENCEPHALIC CYST PRESENTING WITH SPASTIC HEMIPLEGIA

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INTRODUCTION:

Porencephaly is an extremely rare neurological disease characterized by the presence of solitary or multiple degenerative cerebrospinal fluid (CSF) cavities within the brain parenchyma². This condition is probably caused by vascular occlusion resulting from an insult during foetal development or an injury occurring later in life. Its diagnosis depends on demonstrating a well-defined CSF-filled space-occupying lesion communicating with ventricles on CT scan or MRI of brain¹. They are often associated with other malformations of the brain, including microcephaly, abnormal patterns of adjacent gyri and encephalocele. Affected infants tend to have many problems including intellectual disability, spastic hemiparesis or quadriparesis, optic atrophy and seizures. Mutations in the COL4A1 and COL4A2 genes have been described in cases of familial porencephaly.¹ The rarity of occurrence and varied presentation of such a lesion present a challenge to clinicians. Little is known about the pathogenesis and appropriate management of porencephalic cysts. The prognosis of porencephaly depends on the location and extent of the cyst. The best way to contain the increase in incidence of congenital porencephaly is genetic counselling and prenatal testing in affected individuals and those at risk.⁷

KEY WORDS: Spasticity, porencephalic cyst, developmental delay.

CASE REPORT:

We hereby present a case of 5y3m old male child third born to a non-consanguineous marriage, presenting with complaint of weakness and inability to use both right upper and lower limb noticed by parents since 6m of age. Mother was a registered for antenatal checks .However, it was a home delivery, baby said to have cried immediately after birth.Pre-lacteal feeds were given for 4 days and then breastfeeding started. There was no history of any birth trauma or seizures in neonatal period or thereafter.

The child was apparently normal till 6months of age after which the mother paucity of movements of upper limb and delayed milestones in all the domains of development followed by weakness in right lower limb. Early hand preference was noticed by the motherThe child presently, is capable of doing his own tasks but uses only the left arm and leg for the purpose, he interacts well with friends and plays in a group. Upon eliciting past history, there was history of difficulty in carrying the child during infancy with no history suggestive of any cranial nerve involvement, visual or auditory disturbances.

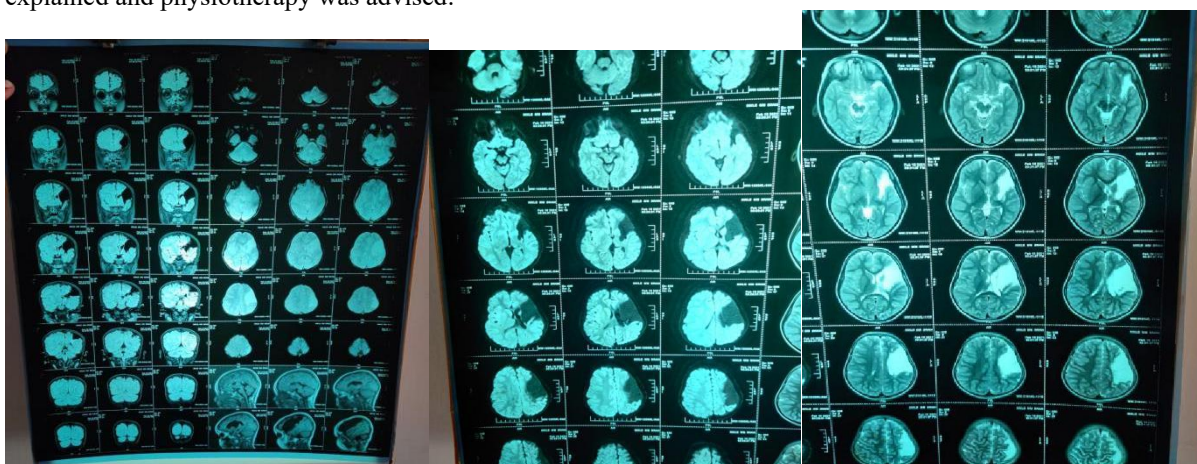
On examination, the child was found to be well thriving. Position of the right upper limbs was observed to be adduction at the shoulder joint, extension at the elbow joint with forearm pronation and flexion of fingers. Hyperextension of right knee joint was also noticed.

Higher mental functions and memory was normal. There was no evidence of cranial nerve involvement. Mild wasting noted in right upper and lower limb, with hypertonia and a power of 4/5. Superficial reflexes were normal, except for plantar, which was extensor on right side and Flexor on left. All the Deep tendon reflexes were exaggerated. Circumduction gait noted. All cortical sensations were intact and No cerebellar signs or signs of meningeal irritation were present.

Examination of other systems was found to be normal.



MRI brain was taken which showed volume loss involving the left cerebral hemisphere, large porencephalic cystic lesion in the left frontoparietal region and ganglio capsular region with gliotic change along the margins, mild ex vacuo dilatation of left lateral ventricle, Wallerian degeneration seen involving the left thalamus and left half of midbrain, pons and medulla, left MCA flow void appears smaller than the right side. Prognosis was explained and physiotherapy was advised.



DISCUSSION:

Porencephaly is an extremely rare neurological disease characterized by the presence of solitary or multiple degenerative cerebrospinal fluid (CSF) cavities within the brain parenchyma.² In this case, the child was noticed to have attained milestones appropriate for age till 6m of age following which there was temporary cessation of milestones and hence delay noted.⁹ The child is presently having difficulty in using his right upper and lower limbs whereas left sided activity is preserved. On neuroimaging, a large porencephalic cyst was noticed probably due to an old infarct most likely in the internal capsule as no cranial nerve is involved, which lead to his current disability.⁸ No evaluation was done at the onset of weakness and the management will be physiotherapy to improve the condition of his life.⁸

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