CONGENITAL BILATERAL PERISYLVIAN SYNDROME: CASE REPORT AND REVIEW OF LITERATURE

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ABSTRACT
Congenital bilateral perisylvian syndrome (CBPS) is a rare structural malformation of the brain in which the underlying anomaly is polymicrogyria. Polymicrogyria is a malformation of cortical development that is characterized by abnormal arrangement and excessive folding of cerebral cortical cell layers, often with fusion of the gyral surfaces, which can be focal or regional or involve the whole cortical mantle. Clinical manifestations depend upon the anatomical region of the brain involved. We report a case of 7-year-old male child with magnetic resonance imaging findings of CBPS.

Keywords:
Congenital bilateral Perisylvian syndrome, epilepsy, magnetic resonance imaging

INTRODUCTION
Congenital bilateral perisylvian syndrome (CBPS) is an extremely rare, late migration disorder of the brain characterized by pseudobulbar palsy, mental retardation, epilepsy and bilateral perisylvian polymicrogyria. This syndrome was originally described by Graff-Radford et al. in identical twins.

CASE REPORT
This was a case report of a 7-year-old male child, with a history of generalized tonic-clonic seizures since 2 years, born of vagina delivery with breech presentation. The child had delayed milestones, poor phonation with drooling of saliva and mental retardation and had feeding and swallowing difficulties with protrusion and movement of the tongue moderately impaired. On examination, there was atrophy of the muscles below knee with exaggerated plantar extensor reflexes on both sides. Reflexes were brisk on both sides. And also had bisyllabal speech (normally seen in 12-month-old infants). Magnetic resonance imaging (MRI) of the brain was performed, which showed the multiple small gyri in bilateral parieto-occipital cortices [Figure 1]. There was thickening of the grey matter around the sylvian fissure bilaterally with irregular cortex and widening of the fissures [Figures 2 and 3]. Bilateral sylvain fissures were hypoplastic and extended dorsally up to the perirolandic region. The body of lateral ventricles shows inverted appearances, typical for this condition.

DISCUSSION
CBPS is a migration disorder of the brain associated with distinctive clinical and imaging features. The clinical spectrum may vary from mild speech difficulties to severe disability, intractable seizures and cognitive and behavioral problems. The exact cause is not known, although bilateral cerebral hypoperfusion, possible injury during the neuronal migration, post-migational vascular accident and gene mutation are the postulated mechanisms. Familial, autosomal and X-linked inheritance has been reported. CBPS has been associated with the chromosomal abnormalities and malformations such as arthrogryposis,[1] clubfeet,[1] micrognathia,[1] polydactyly, constriction band syndrome[2] and pituitary hypoplasia.[3] Our case had hypotonia with no pyramidal signs. He had restricted tongue movements, drooling of saliva, feeding and swallowing problems and lack of speech and language development.

Seizures in CBPS usually begin between the ages of 4-12 years and are poorly controlled in about 60% of patients. Epilepsy was found in almost 90% of cases in the series reported by Kuzniecky et al.[1,4] Epileptic spectrum in this syndrome is broad with seizures presenting as infantile spasms,
generalized tonic-clonic, typical and atypical absences, drop attacks progressing to Lennox-Gastaut syndrome. Our child presented with generalized tonic-clonic seizures at age of 5 years. He had epileptiform discharges with alpha rhythm (8-9 Hz) symmetrically in bilateral centro-temporal region on awake electroencephalogram (EEG) and multiple seizures on a daily basis, ranging from absences to generalized tonic-clonic seizures, which was resistant to medical treatment with anti-epileptic drugs.

CBPS has been classified on the basis of their predominant distribution. The presentation of patients depends upon the distribution. Bilateral frontal polymicrogyria is associated with the developmental delay, mild spastic quadriplegia, impaired language development and epilepsy.

Essential criteria seen in about 100% of cases are oropharyngoglossal dysfunction, moderate to severe dysarthria and bilateral Perisylvian malformations on imaging. The additional criteria seen in nearly 85% of cases are mental retardation, delayed milestones, epilepsy and abnormal EEG. Majority of cases of CBPS have developmental, cognitive, behavioral, speech and language difficulties and epilepsy. Seizures are difficult to treat in the majority and resistant to antiepileptic medications, as in our case. Prognosis for epilepsy cannot be predicted based on the early response to treatment. Callosotomy has been a valuable treatment strategy in those with intractable drop attacks. The presence of esophageal malformations and chromosomal abnormalities and other malformations may be associated with a poor prognosis.

Antenatal diagnosis using ultrasound can be difficult as the regions of the brain that are involved in this malformation may not have reached their final folding until birth. Computed tomography scan may not always identify the polymicrogyria. Prenatal diagnosis using fetal ultrasound and MRI may be difficult as the regions of the brain involved in this malformation may not have reached the final folding before birth. However, few studies have identified bilateral polymicrogyria by prenatal MRI. Hence, MRI is the investigation of choice for detection of this condition.

CONCLUSION

CBPS is a late migration disorder of the brain, which is recognizable by MRI brain and should be suspected clinically in any infant or child presenting with oromotor dysfunction, pseudo-bulbar signs, developmental delay and seizures.

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