25th Annual Conference of INDIAN SOCIETY OF NEURORADIOLOGY

Title of the E-Poster : CONGENITAL BILATERAL PERISYLVIAN SYNDROME: A RARE CASE

ISNR 2024 HYDERABAD 8th 9th 10th March

Indian Society of Neuroradiology

PRESENTATION

Presenting Author's Name & Affiliation: DR.R.SAI KUMAR, OSMANIA MEDICAL COLLEGE

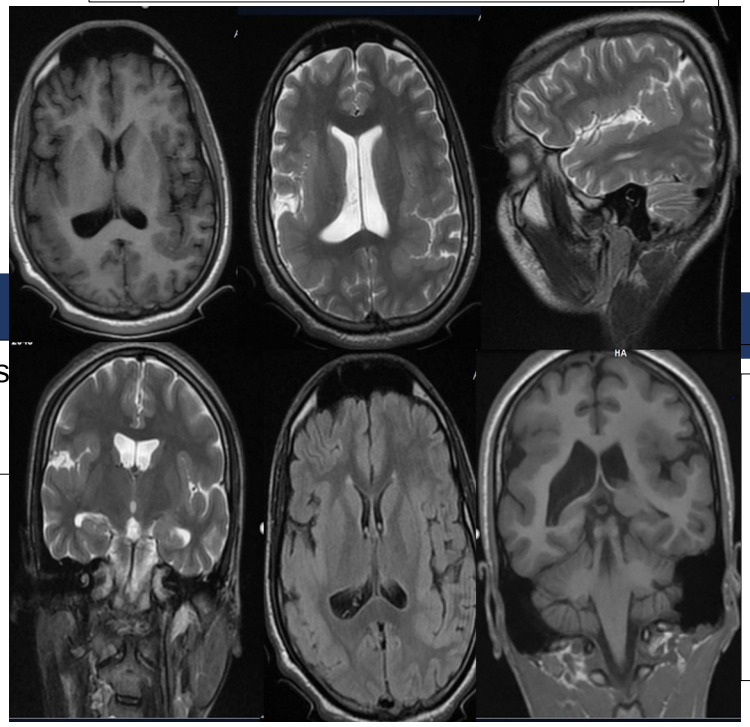
Co - Author's Name & Affiliation :DR SUNITHA RAJA J(PROF & HOD RADIOLOGY) DR NADEEM AHMED (PROF), OSMANIA MEDICAL COLLEGE

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[1] in identical twins.

MATERIALS / METHODS

18year old male patient, Informant being mother presented with a history of generalized tonic clonic seizures, born of vaginal delivery with breech presentation. The child had delayed milestones, mental retardation and had difficulty in speaking, difficulty in chewing since childhood and Unable to walk. On examination, there was atrophy of the muscles below knee with exaggerated plantar extensor reflexes on both sides. MRI brain done using GE 1.5 T machine.



AIMS / OBJECTIVES

To describe the Neuroimaging findings in congenital bilateral perisylvian syndrome.

RESULTS & DISCUSSION

IMAGING FINDINGS :On T1 W Axial Images there is relatively symmetrical thickened insular cortex. On T2W axial images there is isointense thickening of insular cortex with blurring of grey white matter junction. On T2W Sagittal section there is cortical thickening noted with extension along insular opercula. On T2W Coronal there is frontotemporal opercular thickening. On FLAIR sequence there is decreased sulcation with insular cortical thickening. On inversion recovery sequences subtle polymicrogyria at insular cortex with temporo-parietal opercular thickening noted.

- **DISCUSSION:** Congenital bilateral perisylvian syndrome includes developmental delay, variable cognitive deficits. Seizures are common. pseudobulbar symptoms (dysphagia , dysarthria) are the direct result of involvement of the insular and opercular regions. Causes are Genetic factors linked with a mutation on the long arm of the X chromosome (Xq27.2-q27.3 and Xq28) , the Perisylvian syndrome may also be acquired for example from a local malfunction in the blood circulation of the brain, or an infection in the relevant area of the temporal lobe during the foetal stage like congenital cytomegalovirus infection. Imaging characteristically reveals perisylvian and perirolandic cortical malformations (polymicrogyria). Polymicrogyria is a post migrational developmental malformation in which neurons reach the final destination in the cortex; However, they have an abnormal distribution. There are multiple abnormally small convolutions with too few sulci. The opercula are dysplastic and incomplete and the Sylvian fissures are wide and underdeveloped[2].

CONCLUSION

Congenital bilateral perisylvian syndrome is a rare neurological disorder associated with pseudobulbar palsy, cognitive deficits, and epilepsy 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.

REFERENCES:

1.Gowda AK, Mane RS, Kumar A. Congenital bilateral perislyvian syndrome: case report and review of literature. J Clin Neonatol. 2013 Oct;2(4):196-8.

2.El Mandour J, Amsiguine N, Isumbo P, El Fenni J, Edderai M. Congenital bilateral perislyvian syndrome: A rare case report. Radiol Case Rep. 2023 Apr 5;18(6):2066-2068.