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Case Report

Radiology Section

Bilateral Perisylvian Syndrome with Typical Imaging Features– A Case Report

RAMA KRISHNA NARRA[1], ANNE MAHESH KRISHNA[2], BODDEPALLI MANJEERA[3], PUTCHA ANUSHA[4]

ABSTRACT

Bilateral perisylvian syndrome is a congenital neurological disorder characterised by abnormal distribution of neurons in cortex. Unilateral entity may also occur but is less common. Bilateral perisylvian syndrome patients present with pseudobulbar palsy, cognitive impairment and seizures. Imaging findings consist of vertically oriented Sylvian fissure continuous with central/post-central sulcus associated with perisylvian polymicrogyria and septo-optic dysplasia. Authors report a case of a 12-year-old patient who presented with neurologic manifestations and diagnosed as bilateral perisylvian syndrome with typical MR imaging findings.

Keywords: Polymicrogyria, Sylvian fissure, Septo-optic dysplasia

CASE REPORT

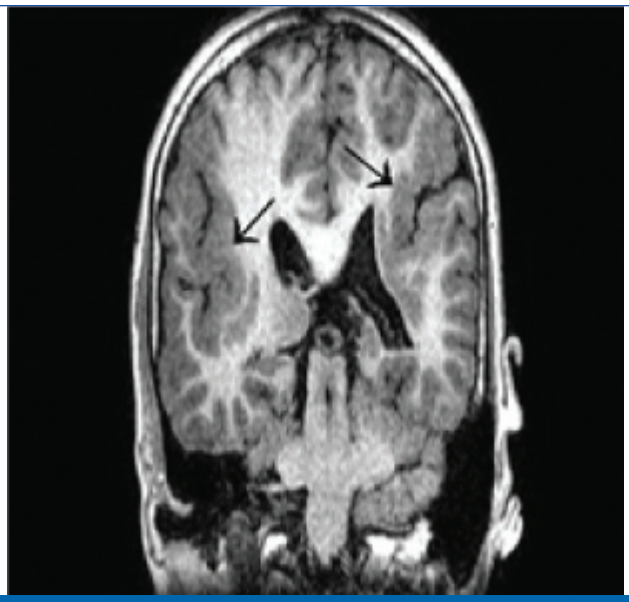
A 12-year-old male patient presented to the Department of Paediatrics with a history of difficulty of speech and complex partial seizures with increasing frequency since 10 years. At five years of age, the patient underwent Computed Tomography scan of the brain and it showed dysplasia of bilateral Sylvian fissures. Then treatment was started with Sodium Valproate (dose was not known) but the patient used the treatment irregularly. Then the patient visited the present hospital. No history of similar complaints in the family members. Mode of delivery of the child was normal vaginal delivery. On examination, general condition of the patient was good and vitals were stable. Blood pressure was 120/80 mmHg; Pulse rate= 76/minute; Respiratory rate= 14/minute; Temperature= 98° F. On systemic examination, impaired mobility of the tongue with dysarthria was noted.

Laboratory investigations included Haemoglobin=14 g/dL (Normal=12-15 g/dL); White blood cells=6.0×10^3/mm^3 (Normal=4.0-10.9x10^3/mm^3); Red blood cells=4.58×10^6/mm^3 (Normal=4.0-5.4x10^6/mm^3); Platelet count=250×10^3/mm^3 (Normal=150-400x10^3/mm^3). Liver function tests including Total bilirubin, Alkaline Phosphatase, Serum Glutamic-Oxaloacetic Transaminase (SGOT), Serum Glutamic Pyruvic Transaminase (SGPT) were normal. Renal function tests including blood urea and serum creatinine were normal. EEG findings were focal sharp and slow waves over the bilateral cerebral hemispheres.

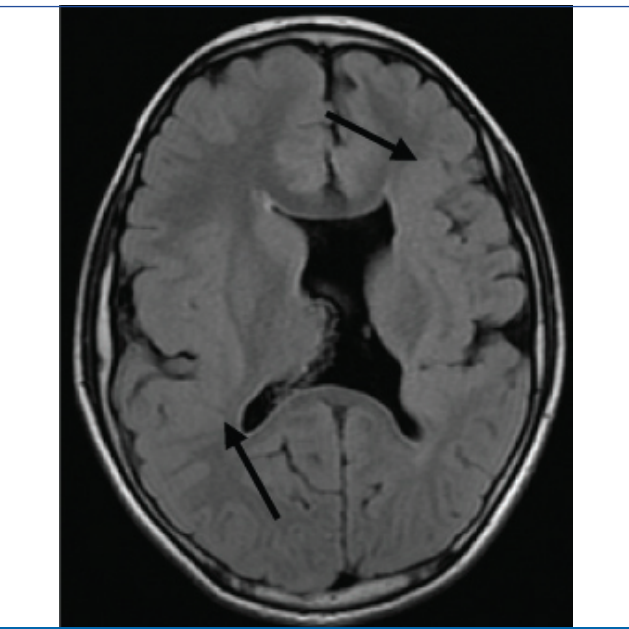
Magnetic resonance imaging of Brain was advised to rule out the possibility of mesial temporal sclerosis. Scan was done using PHILIPS Achieva 1.5T MRI scanner. Sequences used are T1W Axial, T2W Axial, FLAIR Axial, DWI, ADC, GRE; FSPGR BRAVO (Fast spoiled gradient echo brain volume) Axial, Sagittal and coronal sections. The findings on MRI included:

- Widened bilateral Sylvian fissures [Table/Fig-1,2] with thickened and irregular cortex, blurring of gray-white matter junction in bilateral perisylvian regions [Table/Fig-3,4] noted–Suggestive of Polymicrogyria.
- Absent septum pellucidum with dilated left lateral ventricle noted [Table/Fig-5].

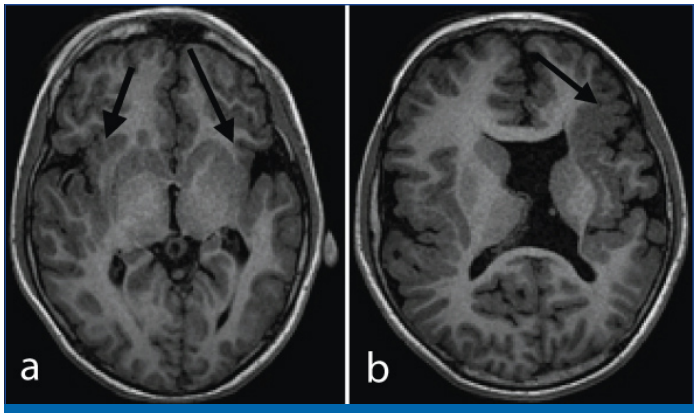
Based on the clinical and typical imaging findings, a diagnosis of bilateral perisylvian syndrome was made. Treatment was initiated with Clonazepam 1.0 mg/day and speech therapy. On follow-up the patient had symptomatic improvement and there was no fresh episode of seizure.



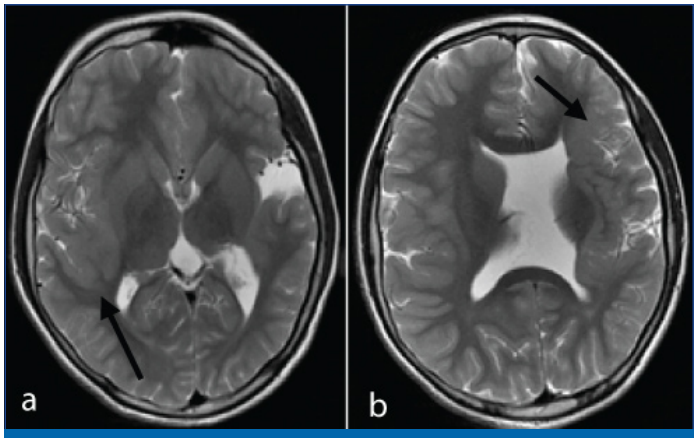
[Table/Fig-1]: Coronal FSPGR BRAVO (Fast spoiled gradient echo brain volume) image showing widened bilateral Sylvian fissures (arrows).



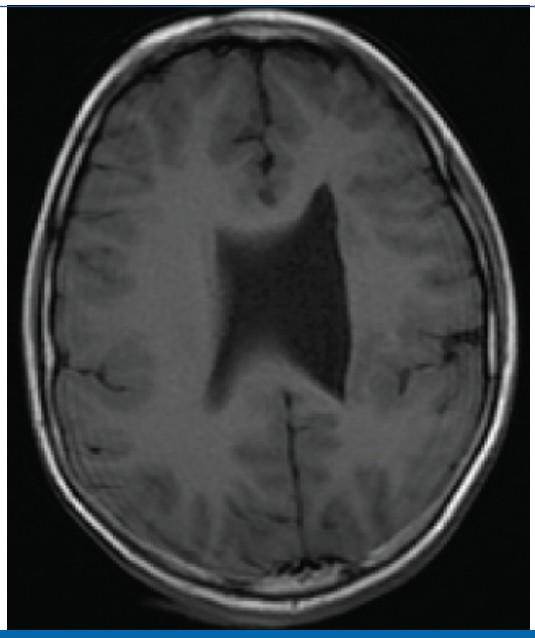
[Table/Fig-2]: Axial FLAIR MR image showing bilateral perisylvian focal cortical thickening (arrows).



[Table/Fig-3]: a) FSPGR BRAVO (Fast spoiled gradient echo brain volume) image depicting widened bilateral Sylvian fissures (arrows); b) FSPGR BRAVO (Fast spoiled gradient echo brain volume) image clearly depicting left perisylvian thickened cortex (arrow) with polymicrogyria.



[Table/Fig-4]: a) Axial T2W MR image showing right perisylvian focal cortical thickening (arrow); b) Axial T2W MR image showing left perisylvian focal cortical thickening (arrow) and absent septum pellucidum.



[Table/Fig-5]: Axial T1W MR image showing absent septum pellucidum and dilated left lateral ventricle.

DISCUSSION

The congenital bilateral perisylvian syndrome is also known as perisylvian polymicrogyria, Worster Drought syndrome and bilateral opercular syndrome [1]. The unilateral perisylvian syndrome is rare compared to its bilateral counterpart [2].

Perisylvian syndrome is a neurological developmental disorder characterised by polymicrogyria in the perisylvian region (i.e., around the Sylvian fissure or lateral sulcus) which is the centre for language and speech in the brain [3].

In a study of 328 patients with polymicrogyria by Leventer RJ et al., a significantly higher prevalence was noted in males compared to females [4]. Perisylvian syndrome can be either familial or acquired. Modes of inheritance in the familial form of the syndrome are X-linked, autosomal dominant and autosomal recessive patterns [1].

Graff-Radford NR et al., in a study described the occurrence of bilateral perisylvian syndrome in identical twins [5]. Acquired form may occur due to infections like cytomegalo virus infection of brain during fetal life. Age at presentation ranges from at birth to 12-years of life. Bilateral perisylvian polymicrogyria is seen in 61% of the cases with polymicrogyria [6].

Clinical presentation in bilateral perisylvian syndrome is with pseudobulbar palsy, whereas in unilateral cases, contralateral hemiparesis is seen. Seizures and cognitive impairment are usually seen in both forms of the disease [2,7]. Abnormalities of cranial nerves will cause partial paralysis of face, jaw, tongue and throat leading to dysphagia, dysarthria and difficulty in mastication. Associated abnormalities in acquiring some developmental milestones will be present leading to certain motor disabilities.

MRI is the imaging modality of choice in perisylvian syndrome. Imaging findings include polymicrogyria cortex with focal cortical thickening, T2W hyper-intensity in adjacent white matter with loss of grey-white matter differentiation in the perisylvian region [8]. Widened and vertically oriented Sylvian fissure which is continuous with central/post central sulcus will be seen. Septo-optic dysplasia may be seen in some cases, which includes absent septum pellucidum, hypoplastic pituitary stalk and hypoplastic optic chiasm/nerves and globes [9].

In the present case, majority of the above-discussed imaging findings and the classic clinical presentation were present, clinching the diagnosis of bilateral perisylvian syndrome.

In a study of 19 patients with frontoparietal polymicrogyria by Chang BS et al., abnormalities in the brainstem, cerebellum and white matter were found to be common [10]. Other diagnostic tools useful in bilateral perisylvian syndrome are Electroencephalography (EEG) and computed tomography scan of the brain.

Diagnosis of bilateral perisylvian syndrome is usually straight forward with few imaging differentials which include isolated polymicrogyria and Lissencephaly Type II.

Bilateral perisylvian syndrome has no cure. Treatment is mainly aimed at control of seizures with antiepileptic medication and speech therapy is also given.

CONCLUSION

MRI is the imaging modality of choice for bilateral perisylvian syndrome. It is essential for early diagnosis and initiation of appropriate management, to prevent progressive clinical deterioration.

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PARTICULARS OF CONTRIBUTORS:
1. Additional Professor, Department of Radiodiagnosis, Katuri Medical College, Guntur, Andhra Pradesh, India.
2. Junior Resident, Department of Radiodiagnosis, Katuri Medical College, Guntur, Andhra Pradesh, India.
3. Junior Resident, Department of Radiodiagnosis, Katuri Medical College, Guntur, Andhra Pradesh, India.
4. Junior Resident, Department of Radiodiagnosis, Katuri Medical College, Guntur, Andhra Pradesh, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Dr. Narra Rama Krishna,
Flat No: 30, 5th Floor, Venkatesh Estate Apartment, ½ Chandramouli Nagar, Guntur, Andhra Pradesh, India.
E-mail: narra.ramki29@gmail.com

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