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Case Report

Radiology Section

Unilateral Perisylvian Syndrome with Subcortical Heterotopia: A Case Report

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ABSTRACT

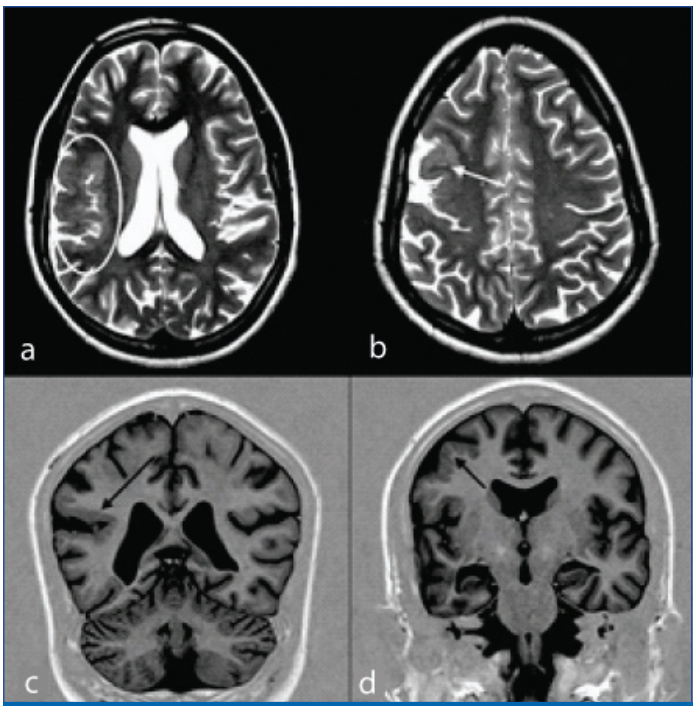
Perisylvian syndrome is a neurological disorder in which a specific area of the brain (called the perisylvian area) develops abnormally, with polymicrogyria as the underlying developmental abnormality. A 21-year-old female reported to the department with complaint of two episodes of seizures per day since six months. Magnetic Resonance Imaging (MRI) revealed multiple small gyri with few sulci noted in the right perisylvian cortex and right frontal lobe, suggesting polymicrogyria. Mild volume loss of right frontoparietal lobes was seen. The small subcortical focus of grey matter signal was noted in the right frontal lobe suggesting of heterotopia. The diagnosis of the perisylvian syndrome is mainly based on the clinical and radiologic features which are treated symptomatically and by multidisciplinary rehabilitative measures as there is no cure for this syndrome.

Keywords: Genetic factors, Polymicrogyria, Seizures

CASE REPORT

A 21-year-old female came with complaints of complex partial seizures with two episodes per day since six months. Contralateral hemiparesis was seen. The patient complained of mild speech developmental issues and certain motor inabilities since her childhood. There were no complaints related to neurological deficits, no fluttering of the eyelids, no generalised stiffening of muscles, no twitching of facial muscles, and no genitourinary system associated complaints. There was no family history record. Occurrence of the malformation among their siblings was not found.

Complete haemogram and blood counts were normal. There was no Electroencephalography (EEG) examination carried out due to patient's unwillingness. MRI was advised to rule out intracranial lesions. Philips Achieva 1.5 T machine was used which revealed multiple small gyri with few sulci noted in the right perisylvian cortex and right frontal lobe suggesting polymicrogyria. Mild volume loss of right frontoparietal lobes was seen. The small subcortical focus of grey matter signal was noted in the right frontal lobe suggesting of heterotopia [Table/Fig-1,2]. Features were suggestive of unilateral perisylvian syndrome with subcortical heterotopia. Treatment was symptomatic and supportive. Anticonvulsant drugs, adrenocorticotropic hormone were used to resolve the seizures. Follow-up of the patient was done upto six months at every one month interval and the dosage of drugs monitored by neurophysician.

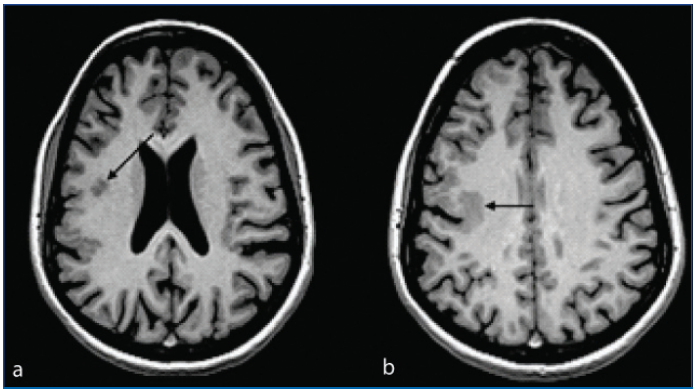


[Table/Fig-1]: Axial T2 images of brain (a and b) showing multiple small crowded gyro in right perisylvian cortex (oval) and right frontal lobe superiorly (white arrow); Coronal T1 inversion recovery images (c and d) showing polymicrogyric cortex in right perisylvian region and frontal lobe (black arrows).

DISCUSSION

The unilateral perisylvian syndrome is rare compared to its bilateral counterpart [1,2]. Clinical work on the subject has not stressed the relevance of unilateral perisylvian dysplasia [1]. However, in one report of 111 patients with congenital hemiplegia studied by CT, nine patients (8%) had brain malformations called "focal pachygyria", seemingly corresponding to unilateral perisylvian dysplasia [3].

Polymicrogyria refers to an abnormal appearance of the cortex with multiple abnormal small convolutions and very few sulci. So, it is an organisation anomaly in which neurons reach their final destination in the cortex but they are distributed abnormally [1,4]. The cortical surface thickness is due to the fusion of adjacent miniature gyri piled on top of one another, and it can be seen in detail. Heterotopia refers to the collection of normal neurons in abnormal locations ranging from the subependymal region to the cerebral cortex.



[Table/Fig-2]: Axial 3D T1 images showing grey matter signal in the subcortical white matter of right frontal lobe (black arrows).

Perisylvian syndrome can be hereditary or acquired. A mutation on the long arm of the X chromosome (Xq27.2-q27.3 and Xq28), which primarily affects male, is important. Perisylvian syndrome can also be caused by a local malfunction in the brain's blood circulation or a similar congenital cytomegalovirus infection in the temporal lobe during the foetal stage. During the fifth or sixth months of pregnancy, the anomaly is usually caused by a post-migration insult. This disease's clinical manifestations include immobility of the tongue and throat, epilepsy, and mild cognitive impairment [5]. In index case, mild volume loss of right frontoparietal lobes was observed. The small subcortical focus of grey matter signal was identified in the right frontal lobe suggesting of heterotopia.

In serious forms neonates may have problems in sucking and swallowing. Dyarthria is characterised by slurred and unclear speech, difficulties forming specific sounds, as well as drooling and swallowing difficulties. Oral motor problems affect the growth of the lower jaw, which may be smaller than normal (micrognathia). Seizures, which can manifest as infantile spasms during the first two years of life and are difficult to treat, are common in the first two years of life. Oropharyngoglossal dysfunction, moderate-severe dysarthria, and unilateral/bilateral perisylvian malformations are essential criteria for diagnosis of this syndrome (i.e., present in nearly 100% of cases). Delayed milestones, epilepsy, mental retardation, and an abnormal electroencephalogram are additional criteria (present in more than 85% of cases). Other criteria for diagnosis (in 50% of cases) include arthrogryposis multiplex, other limb malformations, and infantile spasms [5].

The unilateral perisylvian syndrome described here is the unilateral counterpart to the "bilateral perisylvian syndrome." Radiological features are similar in this syndrome, but lesions cause a distinct clinical picture dominated by suprabulbar signs and more severe epilepsy. Septooptic dysplasia was observed in present case. But, some cases of bilateral perisylvian dysplasia were asymmetric,' suggesting a continuity between the uni and bilateral syndromes [5].

The diagnosis of this disease is based on clinical symptoms, findings of CT, and MRI. In MRI imaging, both morphology and signal intensity observed as abnormal with focal cortical thickening. Image diagnosed the perisylvian and perirolandic cortical malformations. Thickening of grey matter intermixed with shallow sulci and broad gyri involving the perisylvian regions extending dorsally up to the perirolandic region with exposure of insula with associated mild volume loss. Cortical venous drainage abnormalities and developmental venous anomalies were frequently observed. The body of the lateral ventricles may appear inverted. The heterotopic grey matter may be seen within the white matter.

The MRI helps in the grading of severity: perisylvian microgyria of grade 1 extending to the frontal (as in the present) or occipital pole, polymicrogyria of grade 2 that extends beyond the perisylvian region but not to either pole, polymicrogyria of the perisylvian region only is grade 3, polymicrogyria of the posterior perisylvian region only is grade 4 [6,7]. Imaging report of the present case is similar to the study conducted by Sébire G et al., [2].

Perisylvian syndrome has no known cure. Early training in oral motor skills and swallowing will be beneficial. In index case, treatment is symptomatic and supportive. Anticonvulsant drugs, adrenocorticotropic hormone were used to resolve the seizures. Exercises to strengthen the tongue muscles and practice closing the mouth will aid in the prevention of lower jaw deformities. Surgery may sometimes be useful to control epilepsy. If it fails, a vagus nerve stimulator will be the alternative [8].

CONCLUSION(S)

The MR imaging results in multiple small gyri with few sulci observed in right perisylvian cortex and right frontal lobe. Mild volume loss of right frontoparietal lobes was observed. The small subcortical focus of grey matter signal observed in the right frontal lobe. The diagnosis of the perisylvian syndrome is mainly based on the clinical and radiologic features which are treated symptomatically and by multidisciplinary rehabilitative measures as there is no cure for this syndrome.

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