Posterquadranic dysplasia (PQD) is a rare cause of pediatric intractable epilepsy. It is a sporadic cortical development malformation that involves the posterior three lobes of a single hemisphere and spares the frontal cortex. Very few cases have been reported in the literature, mostly as anecdotal reports or as part of large series of refractory epilepsy. It is essential to know about this lesser known entity and differentiate it from other more common similar anomalies such as multilobar cortical dysplasia and hemimegalencephaly as new motor-sparing neurosurgical disconnective procedures have led to dramatically reduced mortality and morbidity rates, apart from gifting the affected children a better quality of life. Magnetic resonance imaging (MRI) is pivotal in astute diagnosis of the condition and accurate delineation of boundaries of the lesion to aid in neurosurgical management. We report one such case of PQD presenting with refractory epilepsy, which was diagnosed on MRI.

**Keywords:** Hemimegalencephaly, magnetic resonance imaging, multilobar cortical dysplasia, posterior quadrant epilepsy, posterior quadrantic dysplasia

**INTRODUCTION**

Malformations of cortical development (MCD) are a well-known cause of drug-resistant pediatric epilepsy. Focal cortical dysplasia (FCD), which represents up to one fifth of MCD, mostly occurs in the frontal and central areas. Cortical dysplasia sometimes extends over multiple cerebral lobes, better known as multilobar cortical dysplasia.[1] Occasionally, this more extensive dysplasia can be seen in the context of hemimegalencephaly, characterized by enlargement of all or parts of a cerebral hemisphere.[1,2] Rarer still, the two processes may affect only the posterior three lobes of one hemisphere, sparing the frontal cortex. This rare entity, which is characterized by a “catastrophic” evolution of refractory epilepsy, is known as posterior quadrantic dysplasia (PQD).[1-3] PQD represents 3–15% of multilobar cortical developmental pathologies.[4] The diagnosis can be accurately made using magnetic resonance imaging (MRI).

It is important to differentiate PQD from other conditions such as multilobar dysplasia and hemimegalencephaly, which have similar radiological appearance. Newer disconnective surgeries that promise a good quality of life are being increasingly suggested for the management of PQD, whereas radical excisions of dysplastic cortex or hemispherectomy may be required for other conditions.

**CASE HISTORY**

A 10-year-old child presented to our hospital with medically refractory epilepsy since birth. Previous records of treatment were unavailable. Presence of a left-sided facial nevus led the pediatrician to refer the case as suspected Sturge-Weber syndrome. There was a history of delayed milestones. Birth history was insignificant. No history of trauma or meningitis was present. On neurological examination, the child was found to have mild mental retardation and minor focal motor deficits. Ophthalmologic evaluation revealed minimal associated visual impairment.

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**Address for correspondence:** Dr. Rohini Gupta Ghasi, Department of Radiodiagnosis, Vardhman Mahavir Medical College and Safdarjung Hospital, New Delhi 110029, India. E-mail: rohini1912@gmail.com

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revealed visual field defects and strabismus. EEG showed epileptiform discharges from left posterior head regions.

Noncontrast-enhanced computed tomography (NCCT) of the brain showed hypertrophy of left temporal, occipital, and parietal lobes with slight diffuse hypodensity in white matter. There was also a focus of coarse calcification in the left occipital white matter. Occipital horn of left lateral ventricle appeared effaced [Figure 1].

MRI of the brain was performed for further characterization. T1-weighted images revealed increased white matter volume in left temporo-parieto-occipital cerebral region causing enlargement of these lobes, with associated heterotopias and polymicrogyria [Figure 2A]. Occipital horn appeared dysmorphic. T2-weighted images depicted hyperintense signal in the periventricular white matter of affected lobes [Figure 2A]. T1 inversion recovery axial images demonstrated polymicrogyria pattern and heterotopias distinctly [Figure 3A and B]. Left frontal lobe appeared normal in morphology and signal intensity. Brain stem, cerebellum, and corpus callosum appeared normal.

On T1-inversion recovery and fluid-attenuated inversion recovery (FLAIR) coronal sequences, the left hippocampus appeared normal in size, though the signal intensity was mildly increased [Figure 4]. T2* gradient echo sequences revealed an area of blooming in left occipital white matter, which corresponded to the areas of calcification noted on the NCCT scan. On the basis of these imaging findings, we arrived at a diagnosis of PQD.

**DISCUSSION**

Major causes of posterior cortex epilepsy include perinatal associated gliosis and malformations such as cortical dysplasia, hemimegalencephaly, and Sturge–Weber syndrome. [5]

FCD represents up to one fifth of MCD, but it is infrequent in parietal and occipital lobes. Posterior cortical involvement may be seen in multilobar cortical dysplasia. Hemimegalencephaly is an anomaly of neuronal migration and proliferation, [4] with excessive growth of a hemisphere, enlargement of ipsilateral ventricle, abnormal gyral pattern, white matter gliosis,
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and abnormal myelination. PQD is a part of this spectrum of MCD where temporal, parietal, and occipital lobes of one cerebral hemisphere are involved. Enlargement of the ipsilateral brain parenchyma occurs due to increased white matter volume but unlike hemimegalencephaly, the frontal lobe is spared.

The overall incidence of PQD is 3–15% of multilobar cortical developmental pathologies. Etiology of this condition is unknown and all reported cases are sporadic. Neuropathologic features of PQD may range from cortical dyslamination without abnormal cellular elements to coexistence of dysmorphic and ectopic neurons and balloon cells of undetermined lineage with abnormal white matter.

Patients present with early onset catastrophic refractory epilepsy. Most common presentation is infantile spasms. Mental retardation, axial hypotonia, and mild contralateral hemiparesis are seen in older children. Developmental delay, minor focal motor deficits, and visual field defects such as visual hemi-inattention and strabismus are common.

Seizure evaluation of patients with PQD using video-EEG demonstrates paroxysmal activities such as multifocal spikes on the posterior regions or diffuse epileptiform abnormalities with posterior predominance. Interictal patterns are characterized by repetitive and rhythmic paroxysmal high-amplitude spikes. Neuroimaging with MRI are the diagnostic modality of choice, which reveal mild-to-moderate enlargement of the posterior quadrant manifested by increased volume of the white matter. The ipsilateral posterior portion of the lateral ventricle shows loss of the normal morphology of trigones and occipital horn. Abnormal high signal in periventricular white matter may be attributed to disordered myelination or gliosis. Gyral abnormalities in the form of simplification of gyral pattern or polymicrogyria are common. Other common findings are ipsilateral hippocampal sclerosis and hypervascularity of the pial structures of the involved region.

It is important to detect this entity early and differentiate it from its counterparts in the spectrum. The conventional treatment for multilobar dysplasia and hemimegalencephaly is functional hemispherectomy. However, hemispherectomy is burdened by higher mortality risk of 2–6% and postoperative motor and neurological deficits, besides longer hospitalization and recovery. The surgical approach for PQD has
evolved toward motor-sparing surgeries such as temporo-parieto-occipital disconnection that aims at isolating the epileptogenic zone and interrupting the discharge pathway.\textsuperscript{[1,7]}

The potential benefits of this approach include smaller craniotomies besides reduction of complications such as blood loss, hemosiderosis, and disturbance of cerebrospinal fluid circulation.\textsuperscript{[8]} Most studies showed promising results in the form of improvement in neuropsychological functioning, resumption of development progress, and improvement in behavior.\textsuperscript{[1,5]}

Early and accurate detection and mapping of abnormality with proper delineation of boundaries using MRI is needed for surgical management to quantify risks of visual, language, and sensorimotor dysfunction.\textsuperscript{[7]}

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Conflict of interest
There are no conflicts of interest.

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