# Sub-Lobar Dysplasia: Neuroimaging and Associated Histopathological Features

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ABSTRACT

Sublobar dysplasia is a rare form of cortical malformation. It has been identified in the literature as having unique imaging findings and histopathological features. We report a rare case of sublobar dysplasia in the right frontal lobe, associated with detailed neuroimaging findings. Histopathological examination ruled out other types of dysplasia, underlying space-occupying lesions, and inflammatory processes.





# CASE REPORT

## CASE REPORT

A 16-year-old boy with epilepsy was transferred to our tertiary hospital with a history of poorly controlled epilepsy for further investigation and management. He was a full-term baby with no history of perinatal complications or prior CNS infections. No family history of epilepsy. He started to have frequent attacks of seizure at age of fifteen. Seizures were generalized tonic-clonic, with associated up-rolling eyes, tongue biting and frothy secretions, followed by post-ictal generalized fatigability headache and sleepiness. No documented Aura. No history of status epilepticus or ICU admission. Electroencephalogram (EEG) showed frequent right frontal polyspike-slow wave. Found to have right frontal lobe lesion, biopsy done and showed sublobar dysplasia. The patient is on Anti-seizure medications including Levetiracetam and Lacosamide, and he is on regular follow up with Neurosurgery and Epilepsy clinic.

**Radiology Description:**

Magnetic resonance imaging (MRI) of the brain revealed an abnormally large well-demarcated area in the right middle and anterior cranial fossa with a diffuse thickened cortex containing thin strips of white matter-like signal intensity (**Figure 1**). The area was undetached from the right inferior frontal gyrus and the hypothalamic structures, which occupied the right suprasellar region, anterior to the enhancing pituitary gland and stalk. It also appeared to be completely detached from the right temporal lobe by the deep sulcus. It measured approximately 4 (CC) × 3.7 (AP) × 5.1 (TS) cm, and it demonstrated iso-signal intensity to gray matter in T1 and T2 / FLAIR. It was not associated with abnormal restricted diffusion, enhancement, or internal magnetic susceptibility changes (**Figure 1**). The lesional area remodeled the adjacent bony structures of the right middle cranial fossa and greater wing of the sphenoid. In addition, it elevated and displaced the right anterior carotid artery and proximal segment of the middle cerebral artery. The pituitary infundibulum deviated to the left side, and there was no midline shift or perilesional edema. There was no direct invasion to the adjacent structures, blood vessels, or bones. There was no intra-orbital extension. The cavernous sinuses and Meckel's cave are intact. Hypoplasia of the corpus callosum was identified in the form of rostral agenesis (**Figure 1**).

**Histopathology Description:**

Histopathological examination showed minimal cortical dyslamination with densely gliotic cerebral and subcortical white matter, highlighted with glial fibrotic acidic protein, and associated with CD34-positive dense vascular spaces. Close focal clustering of neurons was visualized in the cerebral cortex and highlighted using Neu-N immunolabelling. No dysplastic neurons, balloon cells, or microglial nodules were observed. No evidence of neoplastic, inflammatory, or infectious processes was found. Neurofilament staining highlighted the rare axonal swelling present in this section. The sections were also stained with $IDH^{R132H}$, P53 and Ki67, and chromogranin, and all showed unremarkable findings.

## DISCUSSION

**Etiology & Causes:**

Sublobar dysplasia (SLD) is a rare, distinct type of malformation of cortical development (MCD) of unknown etiology, characterized by large, convoluted ribbons of heterotopic grey matter of the brain that has been rarely reported in patients worldwide [**1,2**]. It causes no neurologic deficits but

ultimately results in epilepsy. On neuroimaging, the disease is localized to one cerebral hemisphere or lobe and detached by one or more deep infoldings, giving the lobe within a lobe appearance. Herein, we report an exceedingly rare case of SLD, highlighting detailed neuroimaging and histopathological findings in comparison with previous studies.

SLD is a very rare cortical malformation characterized by a dysmorphic brain region within another normal-appearing cerebral hemisphere. It was first described by Barkovich and Peacock in 1998, who believed it to be a localized variant of hemimegaloencephaly [**1,2**]. Due to its similarity to the localized variant of megalencephaly, SLD has been reclassified under group II C (neuronal migration abnormalities) of the new developmental classification of MCDs and removed from group IV (other unclassified malformations) [**3**]. SLD appears to be a distinct cortical malformation of unknown etiology that causes no neurologic deficits but ultimately results in epilepsy [**1**]. However, possible causes might include abnormal stem cell proliferation and in utero injury [**1**].

**Imaging Findings:**

Neuroimaging findings of SLD in the literature were localized either to one cerebral hemisphere or cortical lobe and more commonly identified in the frontal lobe [**1**,**3**]. Brain MRI demonstrates cortical thickening with reduced gyrification and abnormal sulcal depth and patterns that are detached from the rest of the affected hemisphere or lobe by deep inward folding of the cortex, giving what is called a lobe within a lobe appearance [**3**]. These diseases are commonly associated with other cerebral malformations, such as midline malformation, callosal anomalies, hypoplasia of the cerebellar vermis, ventricular dysmorphism, and venous malformation [**1**,**3**].

Similar findings were found in our case, which demonstrated a lesion mimicking an accessory lobule at the right inferior frontal surface of the brain with a thickened cortex that was detached from the right temporal lobe by a deep sulcus. An atrophic rostral part of the corpus callosum was also observed.

**Differential Diagnoses:**

The histopathological findings reported in SLD include cortical dyslamination, subcortical and leptomeningeal heterotopia, and dysmorphic neurons, correlated to the abnormalities seen in every stage of neuronal development [**2, 3**]. A recent study by Tuxhorn et al. (2009) reported similar histological findings of marked cortical and subcortical gliosis but without dysmorphic cells [**4**]. Subpial gliosis and increased vascularity in the gray and white matter are additional findings observed in SLD [**3**]. Cortical dyslamination, gliosis, dense cortical vascularity, and scattered neuronal clusters were observed. No neuronal dysmorphism was observed. The histological findings helped exclude other types of dysplasia and underlying neoplastic, inflammatory, or infectious processes.

The overall features, along with neuroimaging findings, were consistent with SLD of the right frontal lobe.

In summary, SLD appears to be a distinct cortical malformation that causes no neurological deficits but ultimately results in epilepsy, and it is imperative to identify this syndrome mostly from neuroimaging findings and histopathological correlation.

## TEACHING POINT

SLD is a distinct cortical malformation of unknown etiology that causes no neurologic deficits but ultimately results in epilepsy. The crucial role of neuroimaging findings and the histopathological correlation in diagnosis.

## AUTHORS' CONTRIBUTIONS:

BB, conceptualization, clinical data, writing and editing
ZB, surgical data, interpretation, writing and editing

## DISCLOSURES

Conflict of interest: none of the authors have any conflicts of interest to disclose.
Ethical statement: we confirm that this report is consistent with ethical guidelines.
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Consent: Yes

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## FIGURES

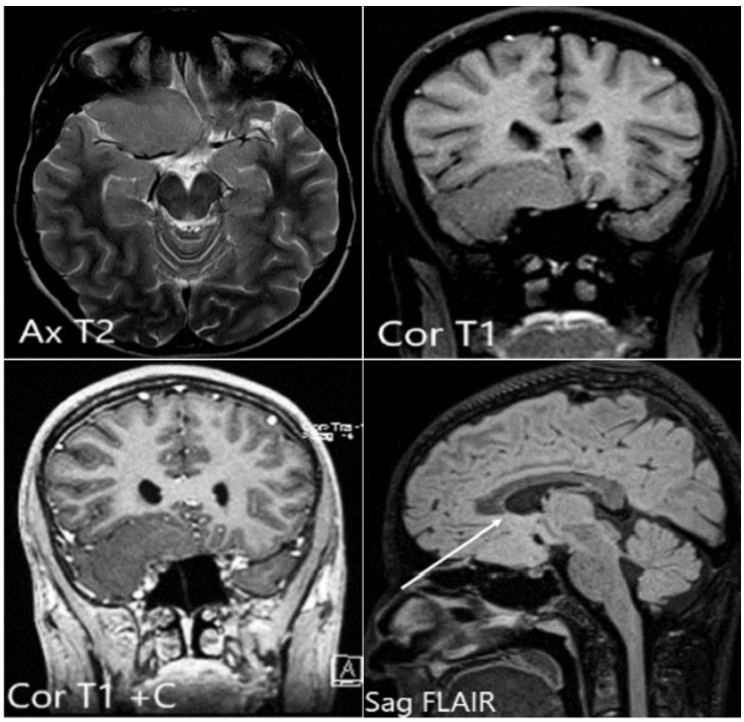


**Figure 1:** A 15-year-old boy, with Sublobar dysplasia.

Technique: Axial T2, Coronal T1, Gadolinium enhanced Coronal T1, Sagittal 3D-FLAR

Findings: Large well-demarcated lesional area in the right middle and anterior cranial fossa, iso-signal to the grey matter, with diffuse thickened cortex containing strips of white matters. The area is undetached from the right inferior frontal gyrus and the hypothalamus, but, separated from the right temporal lobe by a deep sulcus. There is hypoplastic corpus callosum (*arrow*).

## SUMMARY TABLE

| | |
|---|---|
| Etiology | • Unknown, possible abnormal stem cell proliferation or in utero injury |
| Incidence | • Exceedingly rare, 8 reported cases |
| Gender ratio | • Unknown |
| Age predilection | • Congenital |
| Treatment | • Surgical resection can provide excellent seizure control. |
| Prognosis | • No neurologic deficits but ultimately results in epilepsy |
| Imaging findings | • Cortical thickening with reduced gyrification, abnormal sulcal depth and pattern<br>• Separated from the remainder of the affected hemisphere by deep infolding(s) of cortex<br>• Lobe within a lobe appearance.<br>• +/- other malformation (Midline/external malformation, callosal and cerebellar dysgenesis, ventricular dysmorphism ) |

KEYWORDS

Sublobar dysplasia; Cortical malformation; Neuroimaging; Right frontal lobe; Histopathology

ABBREVIATIONS

MRI = Magnetic resonance imaging
AP = Anteroposterior
CC = Craniocaudal
TS = Transverse Section
CD34 = Cluster of Differentiation 34
IDH = Isocitrate Dehydrogenase
P53 = Tumor Protein 53
Ki67 = Marker of proliferation
SLD = Sublobar Dysplasia
MCD = Malformation of Cortical Development

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