

Teaching NeuroImages: Ohtahara Syndrome due to Unilateral Perisylvian Polymicrogyria

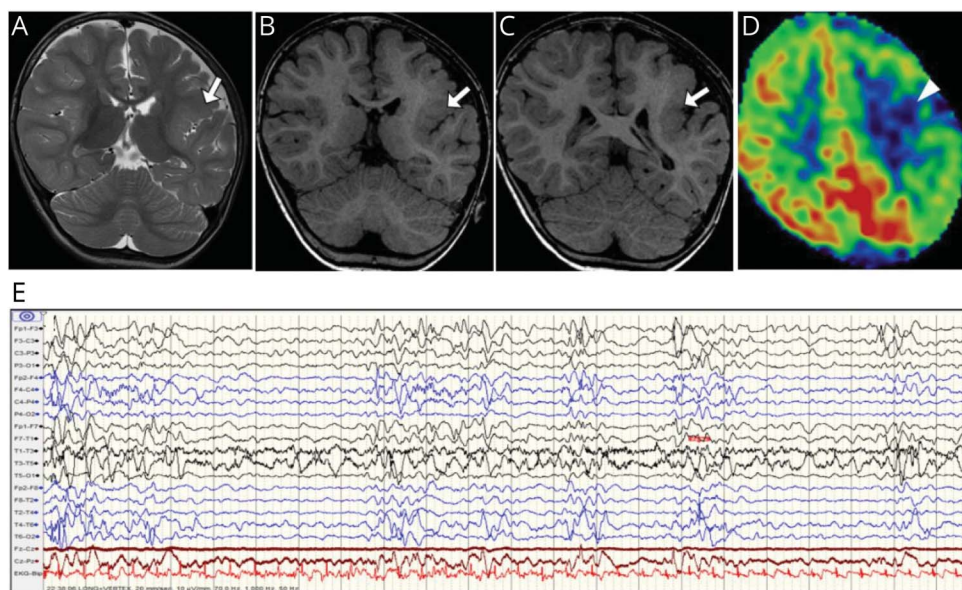
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Figure MRI and EEG



Coronal T2 (A) and coronal T1 (B, C) images show left perisylvian polymicrogyria (A), abnormally thick and bumpy coarse appearance of cortex (B), and widening of sylvian fissure (C). (D) Arterial spin labeling (postlabeling delay 2,500 ms) image shows left perisylvian hypoperfusion corresponding to polymicrogyria (arrowhead). (E) EEG shows typical burst-suppression pattern (time base: 20 mm/s, gain: 10 μ V/mm, high-frequency filter: 70 Hz, low-frequency filter: 1 Hz).

A 10-month-old boy, born of nonconsanguineous parents, without any perinatal insult, presented with tonic spasms from the 13th day of life. He started having tonic seizures from 4 months of age, with global developmental delay. On examination, there were no neurocutaneous markers, facial dysmorphism, or focal neurologic deficits. EEG (figure) showed generalized and multifocal epileptiform discharges along with burst-suppression pattern. MRI of brain revealed left perisylvian polymicrogyria (figure).

Ohtahara syndrome is an electroclinical syndrome, characterized by infantile-onset epileptic encephalopathy and typical burst-suppression pattern on EEG, which remains unchanged during sleep and wakefulness.¹ It is most commonly associated with structural malformations including neuronal migration disorder or dysgenesis, mutations in genes including *ARX*, *CDKL5*, *SLC25A22*, *STXBP1*, and *KCNQ2*, and various metabolic disorders.^{1,2} Presence of structural malformations, like left-sided perisylvian polymicrogyria in our case, precludes

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extensive genetic and metabolic analysis and differentiates it from early myoclonic encephalopathy.

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Name	Location	Contribution
Sabarish Sekar, MD, DNB	Department of Imaging Sciences and Intervention Radiology, Sree Chitra Tirunal Institute for Medical Sciences and Technology, Thiruvananthapuram, Kerala, India	Major role in acquisition of data, study concept, design and drafting of the manuscript
Pavankumar Rudrabhatla, MD	Department of Neurology, Sree Chitra Tirunal Institute for Medical Sciences and Technology, Thiruvananthapuram, Kerala, India	Major role in acquisition of data and EEG discussion

Appendix *(continued)*

Name	Location	Contribution
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Chandrasekharan Kesavadas, MD	Department of Imaging Sciences and Intervention Radiology, Sree Chitra Tirunal Institute for Medical Sciences and Technology, Thiruvananthapuram, Kerala, India	Critical revision of manuscript

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