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Teaching NeuroImage: Clastrum Sign in Febrile Infection-Related Epilepsy Syndrome

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A 40-year-old woman presented with acute encephalopathy and super-refractory status epilepticus six days after a febrile illness. An extensive diagnostic work-up was negative. EEG and brain MRI showed right-predominant abnormalities, including caudate T2/FLAIR hyperintensity, the so-called caudate sign (Figures 1-2). This finding has been described in patients with febrile infection-related epilepsy syndrome (FIRES),¹ a subcategory of new-onset refractory status epilepticus (NORSE) triggered by cytokine storm.² As the caudate sign has been reported also in other cytokine storm-associated disorders, including acute necrotizing encephalopathy, COVID-19-related encephalopathy, and immune effector cell-associated neurotoxicity syndrome (ICANS),³ it may represent a specific marker of cytokine-mediated neuroinflammation.

Figure legend

Figure 1: MRI (FLAIR; A, axial; B, coronal) performed 30 days after the onset of status epilepticus showed right-predominant hyperintensity and swelling of the caudate, putamen and hippocampus.

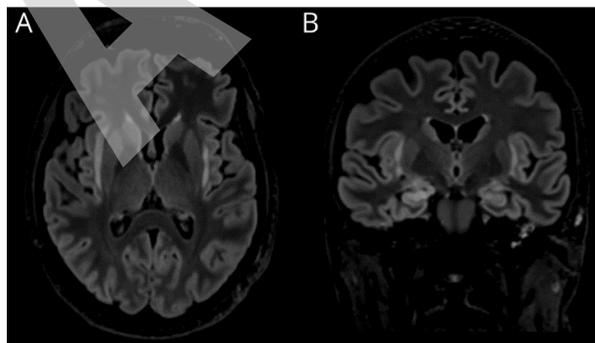


Figure 2: EEG showed bilateral asymmetric lateralized periodic discharges, predominant in the right fronto-temporal region.



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