

Claustrum Sign in Febrile Infection-Related Epilepsy Syndrome (FIRES)

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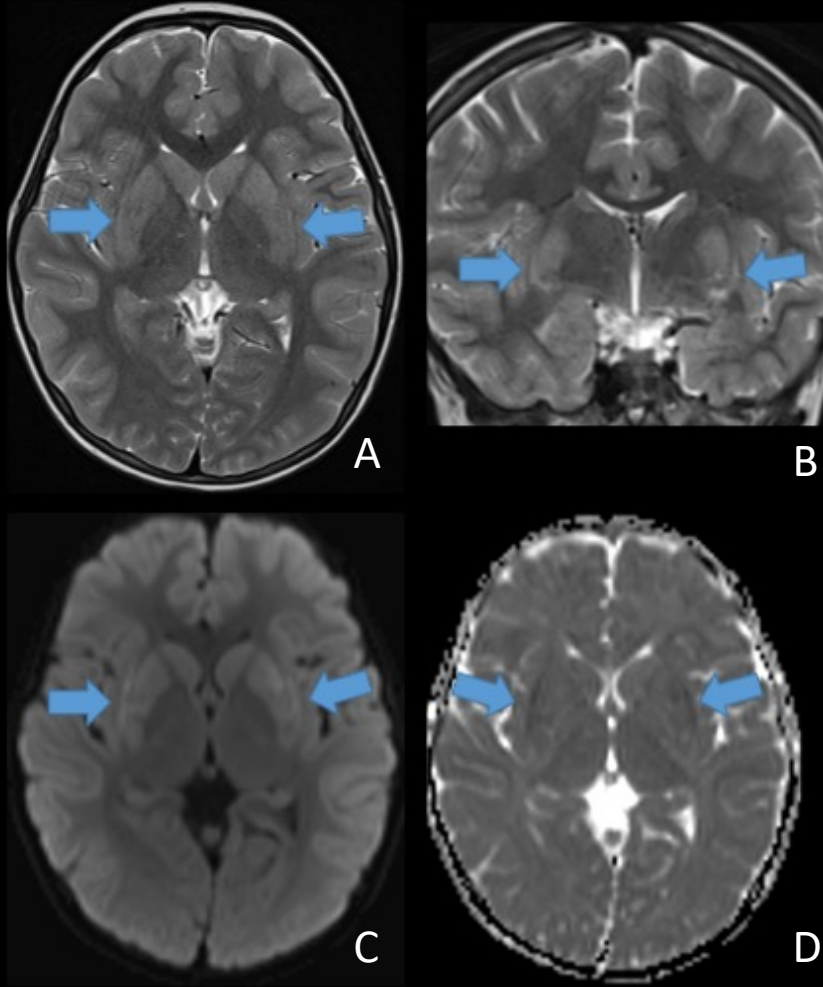


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Clinical Presentation

- Eight year old boy with no past medical history, presented to ED with a generalized tonic clonic seizure lasted about 1 minute.
- He has had a fever 1 week ago with a rash prior to the ED presentation which has been resolved.

Imaging Discussion



T2WI axial (A) and coronal (B) show linear T2 hyperintense signal in bilateral external capsule/clastrum. There is associated diffusion restriction on DWI (C) and ADC (D).

Management and Outcome

- Patient was diagnosed with FIRES (Febrile infection–related epilepsy syndrome). Antiepileptic therapy oxcarbazepine and levetiracetam has been started.

Take Home Points

FIRES begins after a mild nonspecific febrile illness in an otherwise healthy school aged kid. Twenty-four hours to two weeks later, seizures begin and quickly become very frequent and worsen, becoming refractory status epilepticus. The exact etiology is not known but underlying autoimmune or inflammatory mechanism have been proposed.

‘Claustrum sign’ is defined as T2/FLAIR signal abnormality in bilateral claustrum. This sign is reported in cases of FIRES (Febrile infection–related epilepsy syndrome), limbic encephalitis, COVID-19-related encephalopathy and acute necrotizing encephalopathy. This sign is thought to be seen in the setting of cytokine-mediated neuro-inflammation.

References

- Guo K, Hong Z. Claustrum sign in febrile infection-related epilepsy syndrome (FIRES). *Neurol Sci.* 2023 Sep;44(9):3357-3359. doi: 10.1007/s10072-023-06887-6. Epub 2023 Jun 5. PMID: 37273001; PMCID: PMC10240444.
- Hon KL, Leung AKC, Torres AR. Febrile Infection-Related Epilepsy Syndrome (FIRES): An Overview of Treatment and Recent Patents. *Recent Pat Inflamm Allergy Drug Discov.* 2018;12(2):128-135. doi: 10.2174/1872213X12666180508122450. PMID: 29745347.