

A 34-year-old man with Slow Periodic Myoclonus

Teaching NeuroImages

Neurology

Resident & Fellow Section

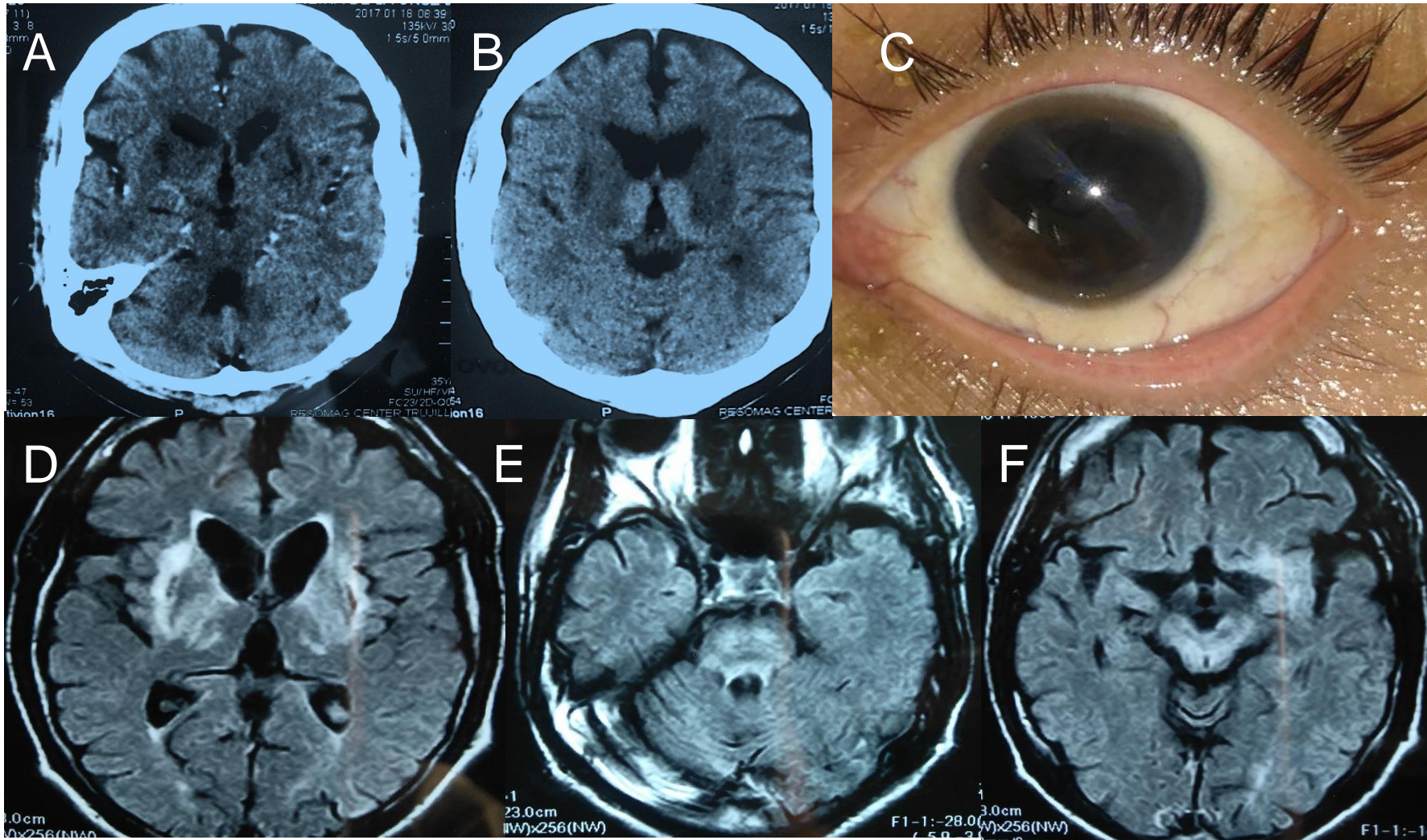
Vignette

- 34-year-old Peruvian man.
- He developed paranoid schizophrenia and, 6 months later, parkinsonism and falls, unresponsive to levodopa, progressing into akinetic mutism with proximal slow periodic myoclonus, synchronous in flexor muscles of both arms and legs (Video 1).
- This case has a similar phenotype to that of a woman patient with visual loss due to chorioretinitis, evolving into a dementia syndrome with slow periodic myoclonus, synchronous in flexor muscles (Video 2).

Imaging



Imaging



Slow Periodic Myoclonus in Fulminant Wilson's disease and Subacute Sclerosing Panencephalitis

- The diagnosis was supported by ocular inspection and neuroimaging features (Figure), as well as by low ceruloplasmin (5.3 U/L), high urinary copper, and diffuse hepatopathy on ecography. He was unresponsive to penicillamine and zinc.
- Slow periodic myoclonus is a well-recognized phenotype of fulminant subacute sclerosing panencephalitis.
- Fulminant Wilson's disease may also rarely evolve into slow periodic myoclonus.
- Slow periodic flexor myoclonus is a manifestation of cortical excitability associated with quasi-periodic, generalized, transient EEG complexes and bears a poor prognosis.