

Brainstem lesion causing paroxysmal ataxia, dysarthria, diplopia and hemifacial spasm (PADDHS)

Enrico Ferrante¹, Maria Raffaella Marazzi²,
Michele Trimboli¹, Davide Dalla Costa³,
Cristina Erminio², Lino Nobili⁴

¹ Neurology Department, AOR San Carlo, Potenza

² Neurosciences Department, Niguarda Cà Granda Hospital, Milan

³ Neurorehabilitation Unit, Niguarda Cà Granda Hospital, Milan

⁴ Neuropsychiatry Unit, IRCCS, Gaslini Institute, Dinogmi, University of Genoa, Italy

Paroxysmal: sudden, stereotyped, 15-30", ~40-times/day attacks.

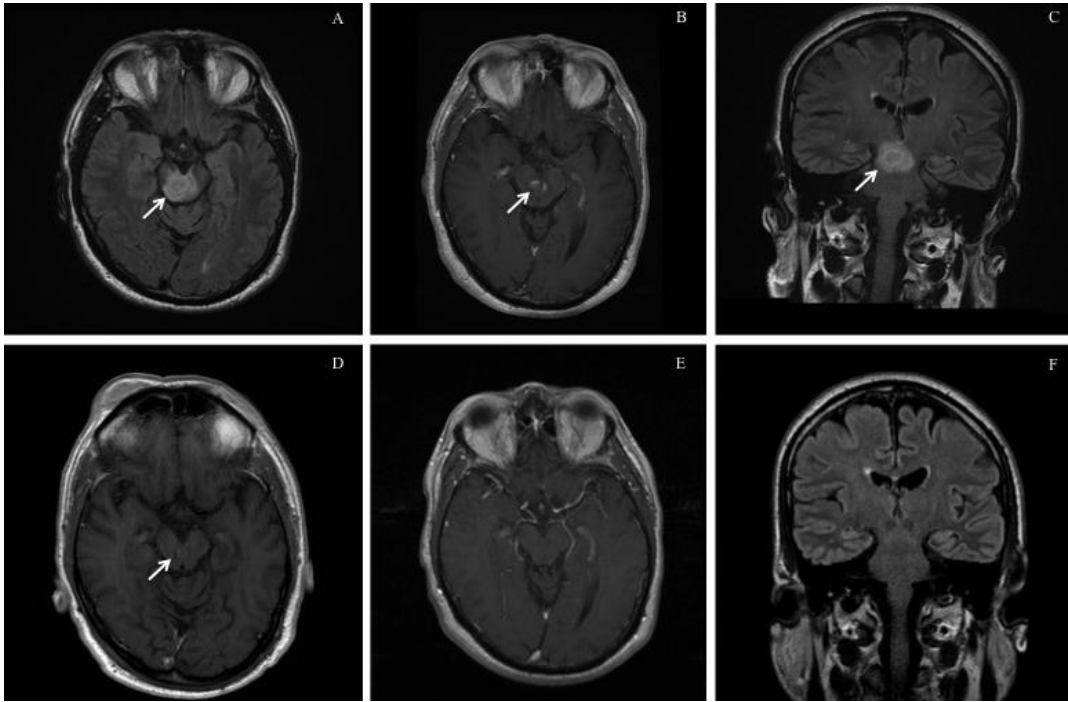
Ataxia: gait ataxia with risk of fall and upper limb ataxia featured by lack of coordination during voluntary movements.

Dysarthria: distorted voice (similar to a 45-rpm record played at 33rpm) .

Diplopia: binocular double vision- two images separated horizontally with a horizontal saccade component.

Hemifacial Spasm: unilateral, tonic, and clonic contractions of the facial muscles.

PADDHS



(A-C) Axial and coronal brain MRI showing enhanced right midbrain and a cystic lesion below the level of the red nucleus, extending to the midbrain-pons junction. After five months, MRI showed only a small level of nuanced midbrain flair hyperintensity (D-F) without enhancement (E). Ictal EEG, during hyperventilation, showed movement artefacts and myogenic potentials without paroxysmal discharges (G).

In this case, despite normal visual acuity and lack of mucocutaneous lesions, **Neuro-Behcet's disease (NBD)** might be a possible diagnosis. Characteristic imaging findings of acute/subacute NBD include *brainstem-centered encephalitis* with contiguous spread into the basal ganglia and subsequent reversibility with small residual central signal changes supporting an oedematous nature of lesions. Moreover, from the clinical point of view, around a third of NBD patients have *single episodes*, a third have repeated relapses with remission, and a third undergo a progressive disease course with accrual of neurological impairments.