Teaching *NeuroImages*: Punctuate and curvilinear enhancement peppering the pons responsive to steroids

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Figure Initial T1-weighted postgadolinium brain MRI

(A) Symmetric enhancement of the pons and both cerebellar peduncles, with pattern highly suggestive of a perivascular distribution. (B) Near complete resolution with corticosteroids. (C) Recurrence of the same lesions without corticosteroids.

A 48-year-old man developed progressive vertigo, ataxia, and dysarthria. MRI demonstrated gadolinium-enhancing abnormalities in the pons

and cerebellar peduncles (figure, A). Further investigations excluded neurosarcoidosis, CNS lymphoma, granulomatosis, and paraneoplastic or

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chronic infectious processes. In particular, 2 CSF examinations revealed mildly elevated protein levels (0.73 g/L, N < 0.40 g/L) without pleocytosis, tumoral cells, or oligoclonal bands.

The patient was treated with IV methylprednisolone (1 g daily for 3 days) followed by oral prednisone (1 mg/kg daily). Symptoms and MRI lesions improved at 3 months, while the patient was receiving 30 mg prednisone daily (figure, B). One month after corticosteroid discontinuation, he experienced a recurrence of the same symptoms. Repeat brain MRI revealed similar lesions (figure, C). Reintroduction of IV methylprednisolone was successful. Chronic therapy with oral prednisone was initiated and he experienced steady improvement. At last follow-up, 4 months after the recurrence, clinical examination only found a right nystagmus while the patient was receiving 25 mg prednisone daily.

The clinical presentation and neuroimaging, and the response to steroids, were highly suggestive of the recently described chronic lymphocytic inflammation with pontine perivascular enhancement responsive to steroids (CLIPPERS) syndrome.¹

MYSTERY CASE RESPONSES The Mystery Case series was initiated by the *Neurology*[®] Resident & Fellow Section to develop the clinical reasoning skills of trainees. Residency programs, medical student preceptors, and individuals were invited to use this Mystery Case as an educational tool. Responses were solicited through a group e-mail sent to the AAN Consortium of Neurology Residents and Fellows and through social media. There were 60 responses to this Mystery Case, 2 of which were submitted via

social media. All responses came from individual residents rather than groups and they were well-reasoned and thoughtful, some of them very detailed with extensive discussion of the differential diagnosis. The majority of respondents (35) identified the particular MRI pattern of enhancement, the clinical signs, and the response to therapy seen in CLIPPERS syndrome. Additional considerations included neurosarcoidosis (8), demyelination (5), Bickerstaff encephalitis (4), Behcet disease (4), Erdheim-Chester disease (1), and intravascular angiocentric lymphoma (1). Two respondents discussed the differential without favoring a particular diagnosis. CLIPPERS appears to be a distinct clinic and radiologic brainstem inflammatory syndrome, though some have raised concerns that radiologically compatible CLIPPERS may conceal a number of different pathologies.²

AUTHOR CONTRIBUTIONS

Dr. Leflauteur: drafting/revising the manuscript, study concept or design. Dr. Bouwryn: drafting/revising the manuscript, study concept or design, analysis or interpretation of data, acquisition of data. Dr. Ahtoy: analysis or interpretation of data, acquisition of data. Dr. Gérardin: analysis or interpretation of data, acquisition of data. Dr. Derrey: drafting/revising the manuscript, acquisition of data. Dr. Maltête: drafting/revising the manuscript, study supervision.

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