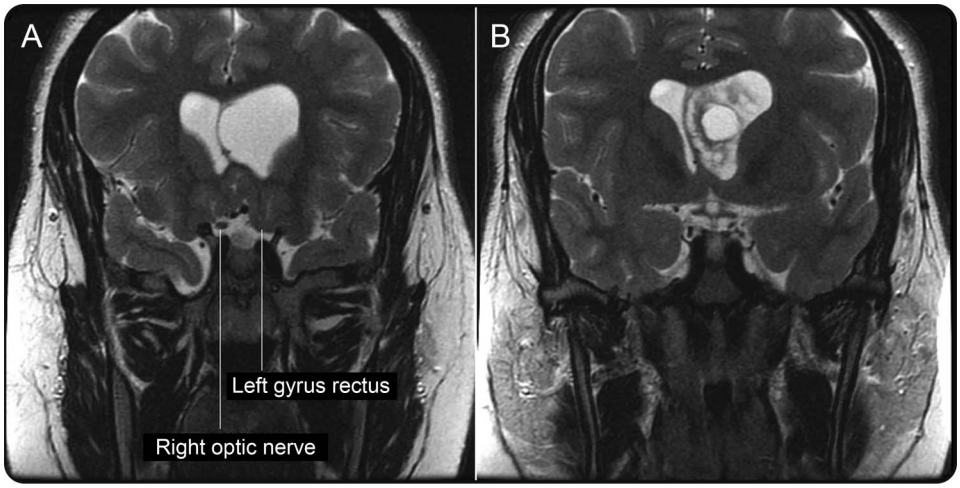
Pseudo-Foster-Kennedy syndrome with optic nerve compression by the gyrus rectus

Figure Optic nerve compression by gyrus rectus and intraventricular mass causing displacement of gyrus rectus



Coronal T2-weighted orbital and brain MRI reveals left lateral ventriculomegaly with downward displacement of the gyrus rectus, resulting in left optic nerve compression (A), due to a left intraventricular mass (B).

A 21-year-old woman presented with headaches and left eye visual loss. Examination revealed acuity 20/20 OD and finger counting OS, a left afferent pupillary defect, papilledema OD, and optic atrophy OS. Left atrophy was unexplained until orbital MRI revealed left nerve compression by the gyrus rectus (figure, A), displaced by an intraventricular central neurocytoma (figure, B). Foster-Kennedy syndrome is characterized by optic atrophy on one side due to direct optic nerve mass lesion compression with contralateral papilledema. This case is termed pseudo-Foster-Kennedy with indirect compressive optic neuropathy due to brain displacement from a tumor distant from the optic nerve.[1]

Ninad Desai, MBBS, Raymund L. Yong, MD, Amish Doshi, MD, Janet C. Rucker, MD

From NYU Langone Medical Center (N.D., J.C.R.); and Mount Sinai Medical Center (R.L.Y., A.D.), New York, NY.

Author contributions: Ninad Desai: manuscript writing and editing. Raymund Yong: conception of idea, manuscript writing and editing. Amish Doshi: manuscript writing and editing. Janet Rucker: conception of idea, manuscript writing and editing.

Study funding: No targeted funding reported.

Disclosure: The authors report no disclosures relevant to the manuscript. Go to Neurology.org for full disclosures.

Correspondence to Dr. Rucker: janet.rucker@nyumc.edu

1. Klingele TG, Gado MH, Burde RM, Coxe WS. Compression of the anterior visual system by the gyrus rectus. J Neurosurg 1981;55:272–275.

Pseudo-Foster-Kennedy syndrome with optic nerve compression by the gyrus rectus

Ninad Desai, Raymund L. Yong, Amish Doshi, et al.

Neurology 2015;85;385
DOI 10.1212/WNL.0000000000001791

an initial brainstem attack sharing chronic lymphocytic inflammation with pontine perivascular enhancement responsive to steroids (CLIPPERS) features. After steroid weaning, MOG-seropositive longitudinally extensive transverse myelitis (LETM) involving the conus appeared, but in absence of brainstem lesions.[1] A diagnosis of CLIPPERS is difficult in this clinical picture.

Brainstem punctate and curvilinear enhancements, a characteristic radiologic finding of CLIPPERS, may conceal several diseases such as glioma, primary CNS lymphoma, lymphomatoid granulomatosis, primary CNS vasculitis, and multiple sclerosis. Except for glioma, all of these diseases initially respond to high doses of steroids, and some could have a relapsing-remitting course in absence of immunosuppressive therapy. However, unlike CLIPPERS, the brainstem is not systematically affected at each relapse and lesion distribution does not remain concentrated in the pons.[2–4]

The Symmonds et al. case highlights a possible pathophysiologic connection between CLIPPERS and demyelinating diseases. Recently, a postmortem analysis performed in a patient sharing all CLIPPERS features revealed the classical perivascular lymphohistiocytic infiltrates but also perivenular demyelinating lesions (as seen in acute disseminated encephalomyelitis).[5] Since perivenular demyelinating lesions were found in only one CLIPPERS patient, it is unlikely that CLIPPERS is a primary demyelinating disease. However, as suggested by the authors, CLIPPERS may induce immunization against MOG antigen.

Author Response: Mkael Symmonds, M. Isabel Leite, Ursula G. Schulz, Oxford, UK: We agree with the important points that Drs. Taieb and Labauge raised in response to our recent report.[1] Our patient presented with clinical and radiologic features consistent with CLIPPERS, even though the subsequent episode of LETM involving the conus had not been previously reported as part of this disease spectrum. While CLIPPERS can relapse, typically with recurrence of brainstem inflammatory features, this does not form key diagnostic criteria and the diagnosis could be consistent with a single episode.

As discussed, the differential diagnosis of CLIPPERS is wide and many of these alternatives will clearly define themselves in time. The histopathologic findings of demyelination in an isolated case at postmortem provides interesting additional support for the hypothesis that CLIPPERS may not be a distinct entity, but rather have a range of underlying etiologies. Our case raises the possibility that anti-MOG antibodies may be causal in some CLIPPERS cases, although we cannot exclude the possibility that CLIPPERS itself causes immunization against MOG epitopes.

1. Symmonds M, Waters PJ, Küker W, et al. Anti-MOG antibodies with longitudinally extensive transverse myelitis preceded by CLIPPERS. Neurology 2015;84:1177–1179.
2. Jones JL, Dean AF, Antoun N, et al. "Radiologically compatible CLIPPERS" may conceal a number of pathologies. Brain 2011;134:e187.
3. Limousin N, Praline J, Motica O, et al. Brain biopsy is required in steroid-resistant patients with chronic lymphocytic inflammation with pontine perivascular enhancement responsive to steroids (CLIPPERS). J Neurooncol 2012; 107:223–224.
4. Ferreira RM, Machado G, Souza AS, et al. CLIPPERS-like MRI findings in a patient with multiple sclerosis. J Neurol Sci 2013;327:61–62.
5. Moreira I, Cruto C, Correia C, et al. Chronic lymphocytic inflammation with pontine perivascular enhancement responsive to steroids (CLIPPERS): postmortem findings. J Neuropathol Exp Neurol 2015;74:186–190.

CORRECTION

Pseudo-Foster-Kennedy syndrome with optic nerve compression by the gyrus rectus

In the Neuro*Image* "Pseudo-Foster-Kennedy syndrome with optic nerve compression by the gyrus rectus" by N. Desai et al. (*Neurology*® 2015;85:385), there is an error in the correspondence address. The note at the bottom should read "Correspondence to Dr. Rucker: janet.rucker@nyumc.org." The authors regret the error.

Author disclosures are available upon request (journal@neurology.org).