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► **To cite this version:**

Zélia Pouillet, Julie Pique, Adil Maarouf, Clemence Boutiere, Audrey Rico, et al. Pure Relapsing Short Myelitis. *Neurology Neuroimmunology & Neuroinflammation*, 2022, 9 (4), 10.1212/uxi.0000000000001167. hal-03812521

**HAL Id: hal-03812521**

**<https://amu.hal.science/hal-03812521>**

Submitted on 12 Oct 2022

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# Pure Relapsing Short Myelitis

## Part of the Multiple Sclerosis Spectrum or New Entity?

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*Neurol Neuroimmunol Neuroinflamm* 2022;9:e1167. doi:10.1212/NXI.0000000000001167

## Abstract

### Background and Objectives

Pure relapsing short myelitis with clinical and paraclinical features suggesting multiple sclerosis (MS) has been described recently. Here, we evaluated the existence of this potential new form of MS by retrospectively searching for similar cases in the databases of the French tertiary MS centers.

### Methods

Patients were included based on the present criteria: at least 2 short (<3 vertebral segments) myelitis episodes; minimum follow-up of 3 years; no MS-like brain lesion during all the follow-up; tested negative for both anti-myelin oligodendrocyte glycoprotein and anti-aquaporin 4 antibodies in serum; presence of oligoclonal bands in CSF; and comprehensive workup to exclude alternative diagnoses.

### Results

Eighteen patients fulfilled all criteria. The sex ratio (females/males) was 5/1; the median (range) age at first relapse was 35.5 (25–54) years, the disease duration was 80.5 (50–308) months, and the annualized relapse rate was 0.36 (0.1–0.5). The median (range) number of relapses per patient was 2 (2–5), and the median (range) Expanded Disability Status Scale score at last follow-up was 1 (0–7.5). In CSF, the median (range) protein level was 0.34 g/L (0.18–0.77), and the median (range) number of mononuclear cells was 3 (0–28). Spinal cord MRI demonstrated a median (range) number of 2 (1–5) lesions per examination and 3 [1–7] on the last examination. Fifty-five percent of lesions involved the cervical levels. Secondary progressive evolution occurred in 3 of 18 (17%) patients.

### Discussion

Pure spinal MS could be a rare entity in the MS disease spectrum. However, the existence of a distinct entity in the inflammatory CNS disorders cannot be excluded.

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Go to [Neurology.org/NN](https://www.neurology.org/NN) for full disclosures. Funding information is provided at the end of the article.

The Article Processing Charge was funded by the authors.

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## Glossary

**anti-AQP4** = anti-aquaporin 4; **anti-MOG** = anti-myelin oligodendrocyte glycoprotein; **ARR** = annualized relapse rate; **DMT** = disease-modifying therapy; **EDSS** = Expanded Disability Status Scale; **MS** = multiple sclerosis; **NOMADMUS** = Neuromyelitis Optica Study Group in France; **OCB** = oligoclonal band.

Cases of pure relapsing short myelitis with clinical and para-clinical features suggesting multiple sclerosis (MS) have been reported recently.<sup>1</sup> Better description of this potential new form of MS is of particular importance because nowadays these patients do not fulfill international diagnostic criteria of MS<sup>2</sup> and could be consequently excluded from effective therapeutic strategies. Here, we evaluated the existence of this potential new form of MS by retrospectively searching for similar cases in the databases of the French centers involved in neuromyelitis optica and associated neurologic disorders (NOMADMUS) network.

## Methods

### Protocol and Participants

To be included, patients had to fulfill the following criteria: age >18 years at inclusion; evidence of at least 2 short (<3 vertebral segments) myelitis episodes; minimum follow-up of 3 years; no typical MS-like brain lesion during all the follow-up; no clinical history or visual evoked potential or eye examination suggesting prior optic neuritis; no history of clinical episode suggesting brain lesion; tested negative for both anti-myelin oligodendrocyte glycoprotein (anti-MOG) and anti-aquaporin 4 (anti-AQP4) antibodies in serum; presence of oligoclonal bands (OCBs) in CSF; and comprehensive workup to exclude alternative diagnoses of myelitis, namely, infections, vascular diseases, and subacute combined degeneration of spinal cord and autoimmune diseases.<sup>3-5</sup> Particularly, other inflammatory causes of myelitis, including sarcoidosis, Behcet disease, paraneoplastic disorders, and connective tissue diseases, were excluded by using biological and imaging explorations. Anonymized centralized (Marseille) reinterpretation of brain MRIs by expert neurologists (consensus required among B.A., J.P., A.M., A.R., and C.B.) was performed to exclude all patients with any typical MS-like brain lesion.<sup>6</sup>

### Standard Protocol Approvals, Registrations, and Patient Consent

The authors obtained ethical approval of national ethical authority (NOMADMUS cohort, CNIL decision DR-2014-558) to conduct the present study. Each participant gave free and informed written consent for anonymized use of clinical, MRI, and biological data for research purposes.

### Data Availability

All data analyzed during this study will be shared anonymized by reasonable request of a qualified investigator to the corresponding author.

## Results

Among 62 patients first screened in the French tertiary MS centers, 18 fulfilled all inclusion criteria (Figure 1).

### Clinical Features

The sex ratio (females/males) was 5/1; the median (range) age at first relapse was 35.5 (25–54) years, the disease duration was 80.5 (50–308) months, and the annualized relapse rate (ARR) 0.36 (0.1–0.5) (Table 1). The median (range) number of relapses per patient was 2 (2–5), and the median (range) Expanded Disability Status Scale (EDSS) score during relapse and at last follow-up was 2.5 (0.5–5.5) and 1 (0–7.5), respectively. Among the 50 relapse cases, 24 (48%) showed pure sensitive signs (paresthesia, numbness, or proprioceptive ataxia); 11 (22%) sensitive and motor signs (arm or lower limb weakness); 7 (14%) sensitive, motor, and sphincter signs; 4 (8%) pure motor signs; 2 (4%) motor and sphincter signs; 1 (2%) sensitive and sphincter signs; and 1 (2%) pure sphincter signs. Of the 50 relapse cases, 30 (60%) involved the 4 limbs and 20 (40%) the lower limbs only.

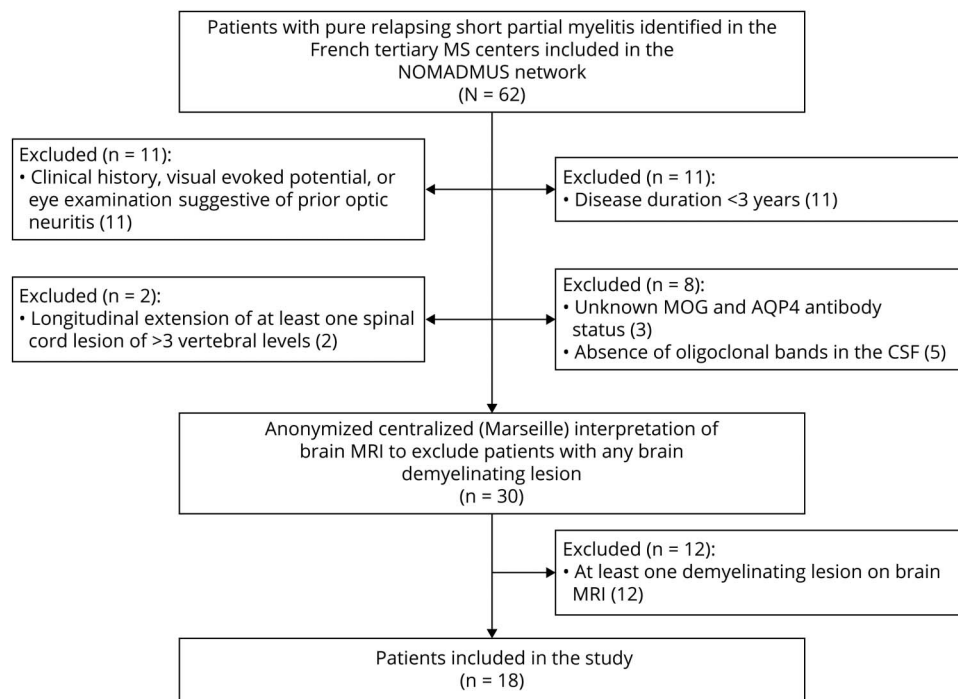
### Laboratory Findings

No patient presented atypical CSF findings for MS. In CSF, the median (range) protein level was 0.34 (0.18–0.77) g/L, and the median (range) number of mononuclear cells was 3 (0–28). Nine of 18 patients were tested at least twice for anti-MOG and anti-AQP4 antibodies, 5 of 9 were tested twice, 3 of 9 three times, and 1 of 9 four times.

### Brain and Spinal Cord MRI

No typical MS-like brain lesion was detected in any patient despite repeat examination (median [range] number of brain MRI examinations per patient 4 [2–9]) (Figure 2). Importantly, 3D fluid-attenuated inversion recovery imaging was available for 15 of 18 patients. A median of 5 (2–6) spinal cord MRI examinations were performed per patient. All spinal cord MRI examinations included sagittal T2/STIR sequences. At least 1 series of axial sequences was available for 8 of 18 patients. Spinal cord MRI demonstrated a median (range) number of 2 (1–5) lesions per examination and a median (range) number of 3 (1–7) lesions on the last examination. The median (range) sagittal extension of the spinal cord lesions was 1 (0.5–2) vertebral segments. Among all spinal cord lesions (n = 67) depicted, 37 (55%) and 30 (45%) involved the cervical and thoracolumbar levels, respectively. Gadolinium injection was performed in 45 examinations, and 21 of 46 (45%) lesions showed gadolinium enhancement. Overall, 18 lesions in 9 patients were explored on the axial plane, and 12 (67%) showed partial myelitis (Figure 3).

**Figure 1** Flowchart of the Patients Included



## Treatment and Progress

In all, 30 of 50 (60%) relapse cases were treated with high-dose IV corticosteroids. The median (range) EDSS score during relapse and after recovery was 2.5 (0.5–5.5) and 1 (0–4.5), respectively.

Disease-modifying therapy (DMT) was used in 12 of 18 (67%) patients (Table 2). In these patients, the median (range) follow-up before and after DMT onset was 55.5 (3–191) and 40 (4–248) months, respectively. The median (range) ARR before and after DMT onset was 0.5 (0.1–1) and 0 (0–0.5), respectively, after excluding patients with a follow-up <6 months before or after DMT onset. Seven of 11 (63%) patients were free from relapse after DMT onset. The median EDSS (range) score at DMT onset and at last follow-up was 2 (0–6) and 2.5 (0–7.5), respectively.

Secondary progressive evolution occurred in 3 of 18 (17%) patients. The median (range) follow-up of these patients was 242 (236–308) vs 64 (36–296) months for other patients. The median (range) EDSS score of these patients was 7 (6–7.5) vs 1.5 (0–4.5) for other patients.

## Discussion

The present study including the French centers involved in the NOMADMUS network reports 18 cases of pure relapsing short myelitis. According to the retrospective design of the study, the number of cases is probably underestimated, which

prevents any conclusion about the prevalence of pure relapsing short myelitis. In addition, some cases were probably not reported because they never experienced a second relapse in that treatment onset occurred just after the first myelitis. However, this therapeutic attitude is highly unusual in France, which limits the number of potential underreported cases.

Because all patients included in the present study had to have OCBs in the CSF, we paid careful attention to searching and excluding all known causes of inflammatory myelitis. Moreover, we also excluded vitamin B12 deficiency in terms of its frequency. However, if a search for other rare metabolic causes of myelopathy such as copper deficiency was not available in the medical chart, patients were not excluded because radiologic and CSF findings did not suggest metabolic disorders.

Several features of these cases argue for the existence of a pure spinal form of MS. First, the characteristics of myelitis highly suggest relapsing MS: no clinical presentation suggested transverse myelitis, clinical presentations mostly suggested an involvement of the posterior part of the spinal cord, spinal cord MRI demonstrating most partial and posterior myelitis in the axial plane. Second, all patients showed typical CSF findings for MS. Third, in all patients receiving MS DMTs, disease activity decreased. Fourth, 17% of patients showed secondary progression—an evolution highly suggestive of MS—several years after disease onset. Finally, we were not able to provide a better explanation than MS in all patients despite extensive explorations. In that way, we recommend

**Table 1** Clinical and MRI Evolution of Patients

	Clinical history	Spinal cord MRI findings	Brain MRI findings <sup>a</sup>
<b>Patient 1, M</b> Age at onset: 35 y Comorbidity: no Family history <sup>b</sup> : no	2011: first relapse (upper limb paresthesia), EDSS score 1, use of HDST 2012: partial recovery, EDSS score 0.5 2017: second relapse (asymmetric lower limb weakness and numbness), EDSS score 3, use of HDST 2018: poor recovery, EDSS score 3 2019: third relapse (asymmetric upper limb weakness), EDSS score 3.5, use of CTS 2020: poor recovery, EDSS score 3.5 2021: stability	2011: C1, C2, and C5-C6 2017: stability 2019: stability 2021: stability	No typical MS-like brain lesion. Controlled in 2011, 2017, 2019, 2020, and 2021.
<b>Patient 2, F</b> Age at onset: 34 y Comorbidity: migraine and 4 miscarriages Family history <sup>b</sup> : no	1996: first relapse (asymmetric lower limb paresthesia), EDSS score 1 1997: good recovery, EDSS score 0 1998: second relapse (asymmetric lower limb weakness and numbness), EDSS score 3, good recovery without HDST 1999: third relapse (lower limb weakness), EDSS score 3, good recovery 2001: fourth relapse (Lhermitte sign and lower limb weakness), EDSS score 4.5 2002: poor recovery, EDSS score 4.5 2003-2020: progressive worsening (severe lower limb weakness and sphincter disorder), last EDSS score 7.5	1998: C3, C4, C5, C6, T1, and T4 2005: stability 2008: stability 2013: +C7 2015: stability 2017: atrophy of the entire spinal cord	No typical MS-like brain lesion. Controlled in 1996, 2001, 2008, 2013, 2015, 2017, and 2018.
<b>Patient 3, F</b> Age at onset: 37 y Comorbidity: rheumatoid polyarthritis Family history <sup>b</sup> : no	1996: first relapse (asymmetric lower and upper limb weakness and sphincter disorder), EDSS score 3 1997: partial recovery, EDSS score 1 2002: stability, EDSS score 1 2007: second relapse (ataxia and asymmetric upper and lower limb weakness), EDSS score 3, use of HDST 2008: partial recovery, EDSS score 2 2013: third relapse (ataxia, asymmetric tetraparesia, and sphincter disorder), EDSS score 5.5, use of HDST 2014: good recovery, EDSS score 2 2016: fourth relapse (asymmetric tetraparesia with numbness and ataxia), EDSS score 5, use of HDST 2017: good recovery, EDSS score 2 2017-2021: stability, EDSS score 2	2008: C6 2012: stability 2016: +C2-C3 and T3 2017: stability 2020: +T5-T6 and T10	No typical MS-like brain lesion. Controlled in 2008, 2012, 2014, 2017, 2018, and 2020.
<b>Patient 4, F</b> Age at onset: 54 y Comorbidity: myeloma Family history <sup>b</sup> : no	2002: first relapse (ataxia), EDSS score 1, no recovery 2007: second relapse (asymmetric lower limb weakness and numbness), EDSS score 3 2008: no recovery (EDSS score 3) 2009-2021: progressive worsening, last EDSS score 6	2017: C3, C5, T2, and L3 2018, 2019, and 2020: stability	No typical MS-like brain lesion. Controlled in 2017, 2019, and 2020.
<b>Patient 5, F</b> Age at onset: 44 y Comorbidity: no Family history <sup>b</sup> : no	2001: first relapse (lower limb paresthesia and asymmetric weakness and saddle hypesthesia), EDSS score 3, use of HDST 2002: partial recovery, EDSS score 1 2005: second relapse (asymmetric lower limb weakness), EDSS score 3, use of HDST 2006: good recovery, EDSS score 1 2008: third relapse (ataxia and lower limb weakness), EDSS score 3, use of HDST 2009: good recovery, EDSS score 1 2011: fourth relapse (ataxia and lower limb weakness and numbness), EDSS score 4, use of HDST 2012: partial recovery, EDSS score 2 2014: fifth relapse (ataxia and lower and upper limb weakness and numbness), EDSS score 5.5, use of HDST 2015: partial recovery, EDSS score 4 2016-2021: progressive worsening with persistence of acute deterioration, last EDSS score 6.5	2001: T7-T8 with gadolinium enhancement 2005, 2010, 2011, 2014, and 2016: stability 2018: + C6-C7 2020: +T4-T5 2018: +C2 2020: stability	No typical MS-like brain lesion. Controlled in 2001, 2003, 2005, 2010, 2011, 2014, 2016, 2018, and 2020.

Continued

**Table 1** Clinical and MRI Evolution of Patients (*continued*)

	Clinical history	Spinal cord MRI findings	Brain MRI findings <sup>a</sup>
<b>Patient 6, F</b> Age at onset: 42 y Comorbidity: 0 Family history <sup>b</sup> : 0	2016: first relapse (4 limb asymmetric paresthesia and Lhermitte sign), EDSS score 2, use of CTS 2017: partial recovery, EDSS score 1 2018: second relapse (4 limb hypesthesia), EDSS score 2.5, use of HDST 2019: no recovery, EDSS score 2.5 2021: stability, EDSS score 2.5	2016: C2-C3 with gadolinium enhancement 2017, 2018, and 2019: stability	No typical MS-like brain lesion. Controlled in 2016, 2018, 2019, and 2020.
<b>Patient 7, F</b> Age at onset: 30 y Comorbidity: no Family history <sup>b</sup> : no	2017: first relapse (4 limb hypesthesia), EDSS score 1 2018: good recovery, EDSS score 0 2020: second relapse (asymmetric hypesthesia), EDSS score 1, use of HDST 2021: good recovery, EDSS score 0	2017: C3, T7, T8, and T9 with gadolinium enhancement of all lesions 2019: stability 2020: +C7-T1 and L1	No typical MS-like brain lesion. Controlled in 2017, 2019, and 2020.
<b>Patient 8, F</b> Age at onset: 25 y Comorbidity: no Family history <sup>b</sup> : no	2015: first relapse (ataxia, Lhermitte sign, and upper limb weakness and numbness), EDSS score 5, use of HDST 2016: good recovery, EDSS score 1 2017: second relapse (lower limb weakness and numbness), EDSS score 2 2018: spontaneous recovery, EDSS score 0 without HDST 2019-2021: stability, EDSS score 0	2015: C2-C3 with gadolinium enhancement 2016: stability 2017: +T3 2020: stability	No typical MS-like brain lesion. Controlled in 2015, 2016, 2017, and 2020.
<b>Patient 9, F</b> Age at onset: 36 y Comorbidity: no Family history <sup>b</sup> : no	2016: first relapse (asymmetric upper limb numbness), EDSS score 1, use of HDST 2017: good recovery, EDSS score 0 2018: second relapse (asymmetric lower limb weakness and numbness), EDSS score 3, use of HDST with good recovery, EDSS score 0 2019: third relapse (Brown-Sequard syndrome), EDSS score 2.5, use of HDST with good recovery, EDSS score 0 2020: fourth relapse (lower limb numbness and paresthesia), EDSS score 2 2021: good recovery, EDSS score 0	2016: C5 2018: +C6 with gadolinium enhancement 2020: +T12-L1 2021: stability	No typical MS-like brain lesion. Controlled in 2016, 2017, 2018, 2019, and 2020.
<b>Patient 10, F</b> Age at onset: 29 y Comorbidity: T8 to L3 fractures <sup>c</sup> Family history <sup>b</sup> : no	2015: first relapse (ataxia and 4 limb numbness and mild weakness), EDSS score 2.5, use of HDST 2016: partial recovery, EDSS score 1.5 2019: second and third relapses (ataxia, 4 limb hypesthesia, and sphincter disorder), EDSS score 4, use of HDST 2020: no recovery, EDSS score 4 2021: stability	2015: C2-C3 and T9 2016: +T3 2017 and 2018: stability 2019: gadolinium enhancement within the C3 lesion 2020: stability and regression of the gadolinium enhancement	No typical MS-like brain lesion. Controlled in 2016, 2017, 2018, 2019, 2020, and 2021.
<b>Patient 11, F</b> Age at onset: 26 y Comorbidity: migraine Family history <sup>b</sup> : migraine	2013: first relapse (hands paresthesia, ataxia), EDSS score 1, spontaneous recovery 2014: second relapse (upper limb paresthesia), EDSS score 1, spontaneous recovery 2016: third relapse (asymmetric 4 limb numbness), EDSS score 2.5, use of HDST 2017: partial recovery, EDSS score 1 2018: fourth relapse (lower limb paresthesia and ataxia), EDSS score 2.5, use of HDST 2019: good recovery, EDSS score 1 2021: stability	2016: C2-C3, C6-C7, and T8-T9 2017: +T10-T11 with gadolinium enhancement 2018: +T6 2020: stability	No typical MS-like brain lesion. Controlled in 2017, 2019, and 2020.
<b>Patient 12, F</b> Age at onset: 62 y Comorbidity: no Family history <sup>b</sup> : no	2015: first relapse (lower limb numbness), EDSS score 0.5 2016: no recovery but stability, EDSS score 0.5 2018: second relapse (asymmetric lower limb weakness), EDSS score 2 2019: no recovery, EDSS score 2 2021: stability	2020: C4-C5, C7, T3, and T6 2021: stability	No typical MS-like brain lesion. Controlled in 2020 and 2021.

Continued

**Table 1** Clinical and MRI Evolution of Patients (*continued*)

	Clinical history	Spinal cord MRI findings	Brain MRI findings <sup>a</sup>
<b>Patient 13, F</b> Age at onset: 28 y Comorbidity: migraine Family history <sup>b</sup> : no	2016: first relapse (4 limb numbness), EDSS score 2, use of HDST 2017: good recovery, EDSS score 0 2018: stability 2020: second relapse (lower limb numbness), EDSS score 2, use of HDST 2021: poor recovery, EDSS score 2	2016: C2, C4, C5, and T3 2018: +C3 2020: +T9-T10 with gadolinium enhancement	No typical MS-like brain lesion. Controlled in 2016, 2018, and 2020.
<b>Patient 14, M</b> Age at onset: 42 y Comorbidity: migraine Family history <sup>b</sup> : no	2017: first relapse (ataxia, asymmetric numbness, and sphincter disorder), EDSS score 3, use of HDST with partial recovery: EDSS score 1 2017: second relapse (4 limb numbness and weakness), EDSS score 2, use of HDST 2018: poor recovery, EDSS score 2 2019-2021: stability, EDSS score 2	2017: C1, C2, C5-C6, T1, T9, and T12 with gadolinium enhancement of all lesions 2018: + C3 with gadolinium enhancement 2019: amelioration 2020: stability	No typical MS-like brain lesion. Controlled in 2017, 2018, 2019, and 2020.
<b>Patient 15, F</b> Age at onset: 34 y Comorbidity: migraine Family history <sup>b</sup> : no	2016: first relapse (sensory deficit), EDSS score 2, use of HDST 2017: good recovery, EDSS score 0 2019: second relapse (sensory deficit), EDSS score 2, use of HDST 2020: complete recovery, EDSS score 0 2021: stability, EDSS score 0	Lack of previous MRI 2020: C6, T6, and T7-T8 2021: stability	No typical MS-like brain lesion. Controlled in 2016, 2017, 2018, 2019, and 2020.
<b>Patient 16, F</b> Age at onset: 39 y Comorbidity: migraine Family history <sup>b</sup> : no	2014: first relapse (lower limb numbness), EDSS score 2 2015: partial recovery, EDSS score 1 2017: second relapse (upper limb numbness), EDSS score 2 2018: partial recovery, EDSS score 1 2019-2021: stability, EDSS score 1	2015: T11 2016: stability 2017: +C3 2019: stability	No typical MS-like brain lesion. Controlled in 2015, 2017, and 2018.
<b>Patient 17, F</b> Age at onset: 33 y Comorbidity: Raynaud phenomenon Family history <sup>b</sup> : no	2014: first relapse (asymmetric paresthesia), EDSS score 1 with spontaneous full recovery: EDSS score 0 2014: second relapse (asymmetric 4 limb numbness), EDSS score 1, full recovery 2015: third relapse (asymmetric 4 limb weakness and numbness and Lhermitte sign), EDSS score 2, use of HDST 2016: good recovery, EDSS score 0 2017-2021: stability, EDSS score 0	2015: C2 with gadolinium enhancement 2016, 2017, 2018, 2019, and 2021: stability	No typical MS-like brain lesion. Controlled in 2015, 2016, 2017, 2018, and 2021.
<b>Patient 18, M</b> Age at onset: 42 y Comorbidity: no Family history <sup>b</sup> : no	2013: first relapse (sphincter disorder), EDSS score unknown 2015: second relapse (4 limb weakness and numbness), EDSS score 4 2016: partial recovery, EDSS score 2.5 2017-2021: clinical improvement with EDSS score 0	2015: C2-C3 with gadolinium enhancement 2016 and 2019: stability	No typical MS-like brain lesion. Controlled in 2015, 2016, and 2019.

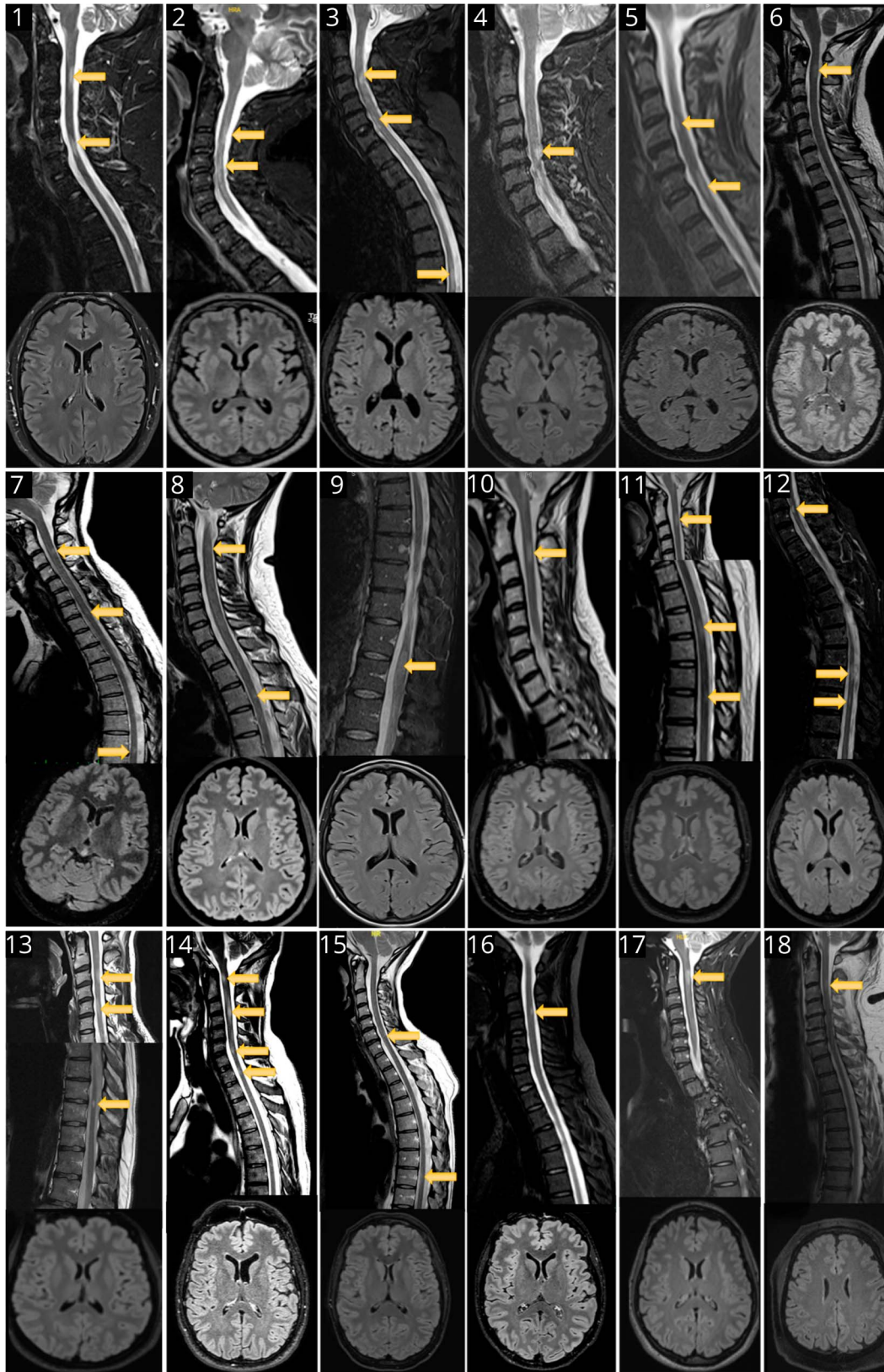
Abbreviations: EDSS = Expanded Disability Status Scale; HDST = high-dose steroid therapy; MS = multiple sclerosis.

<sup>a</sup> Anonymized centralized (Marseille) interpretation of brain MRI by expert neurologists (B.A., J.P., A.M., A.R., and C.B.) to rule out patients with any typical MS-like brain lesion.

<sup>b</sup> Family history: first-degree family history.

<sup>c</sup> Patient 10 had a history of T8-L3 vertebral fractures.

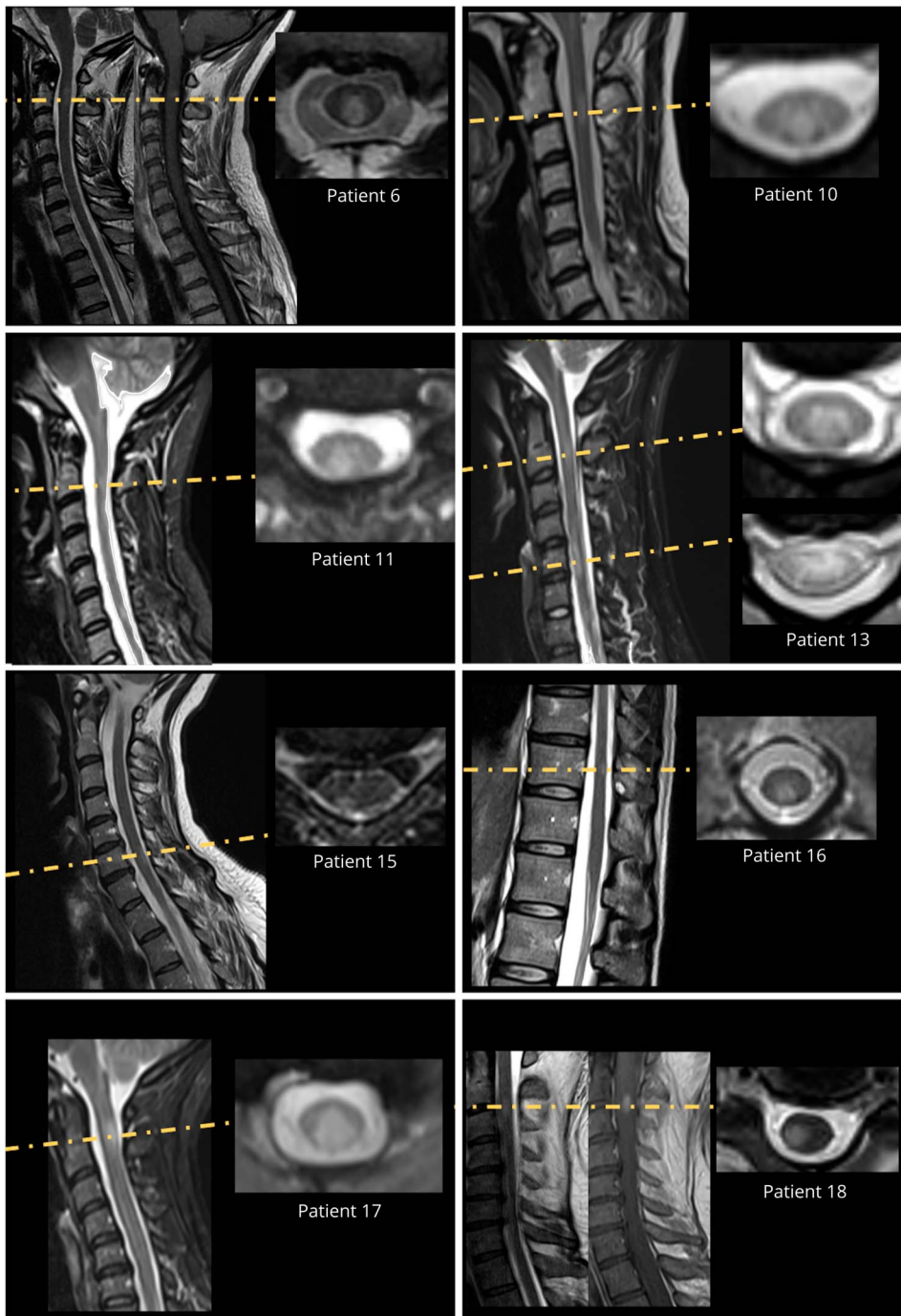
**Figure 2** Last Brain and Spinal Cord MRI of the Patients



The patient's number is displayed in each image.



**Figure 3** Axial View of Spinal Cord Lesion Performed in 8 of 18 Patients



preferentially using DMTs with demonstrated efficacy in MS to treat pure relapsing short myelitis.

Nevertheless, several features may argue for the existence of a possible distinct inflammatory entity. First, the sex ratio was more imbalanced in favor of females as compared with MS. Second, it is unexpected in a pathologic perspective that brain involvement could be totally absent after several years of evolution with MS.

Whatever the nosological classification, the existence of patients with pure relapsing short myelitis argues for systematically adding spinal cord MRI to brain MRI for the imaging surveillance of patients followed after an isolated myelitis episode with OCBs in the CSF. For patients with recurrent myelitis, we recommend performing imaging at least annually as recommended for MS but systematically adding spinal cord imaging to brain imaging. According to the relative low disease activity evidenced in the patients reported here, we do

**Table 2** Characteristics and Evolution of Treated Patients (n = 12)

	Disease duration at DMT onset (mo)	Type of DMT	Mean follow-up after DMT onset (mo)	ARR <sup>a</sup> before DMT	ARR <sup>a</sup> after DMT	EDSS score at DMT onset	EDSS score at last follow-up
<b>Patient 1, M</b> Age at onset: 35 y	108	Fingolimod: 01/20–	18	0.33	0	3.5	3.5
<b>Patient 2, F</b> Age at onset: 34 y	60	Azathioprine: 09/01–01/02 Cyclophosphamide: 01/02–08/02 Mitoxantrone: 2002–2006 Cyclophosphamide: 09/08–2012 Rituximab: 04/16–05/16 Fingolimod: 01/17–	247	0.6	0	6	7.5
<b>Patient 4, F</b> Age at onset: 54 y	191	Dimethyl fumarate: 12/17–06/18 Rituximab: 08/18–	44	0.13	0	6	6
<b>Patient 5, F</b> Age at onset: 44 y	72	Glatiramer acetate: 2007–2012 Fingolimod: 2012–2016 Rituximab: 03/17–	175	0.33	0.21	2	7
<b>Patient 6, F</b> Age at onset: 42 y	24	Teriflunomide: 11/18–	33	1	0	2.5	2.5
<b>Patient 10, F</b> Age at onset: 29 y	40	Interferon beta: 02/19–09/19 Dimethyl fumarate: 09/19–08/20 Natalizumab: 08/20–	30	0.33	0	2.5	4.5
<b>Patient 11, F</b> Age at onset: 26 y	60	Interferon beta: 01/18–	43	0.6	0	1	0.5
<b>Patient 13, F</b> Age at onset: 28 y	56	Azathioprine: 10/20–	9	0.5	0	2	2
<b>Patient 14, M</b> Age at onset: 42 y	3	Mycophenolate mofetil: 10/17–	45	NA <sup>a</sup>	0	2	2
<b>Patient 15, F</b> Age at onset: 34 y	55	Mycophenolate mofetil: 05/21–	3	0.5	NA <sup>a</sup>	0	0
<b>Patient 16, F</b> Age at onset: 39 y	44	Mycophenolate mofetil: 08/18–	35	0.66	0	1	1
<b>Patient 18, M</b> Age at onset: 42 y	42	Interferon beta: 12/16–09/17 Dimethyl fumarate: 09/17–	55	0.66	0.5	0	0

Abbreviations: ARR: annualized relapse rate; DMT: disease-modifying therapy; EDSS: Expanded Disability Status Scale.

<sup>a</sup> Patients with a follow-up < 6 months before or after DMT onset were excluded from the analysis.

not recommend exceeding annual imaging in the absence of relapse or therapeutic considerations.

Pure spinal MS could be a rare entity in the MS disease spectrum. However, the existence of a distinct entity in the inflammatory CNS disorders cannot be excluded. Future studies are needed to disentangle these 2 interpretations.

## Study Funding

No targeted funding reported.

## Disclosure

Dr. Pouillet reports no disclosures relevant to the manuscript. Go to [Neurology.org/NN](https://www.neurology.org/NN) for full disclosures.

## Publication History

Received by *Neurology: Neuroimmunology & Neuroinflammation* December 6, 2021. Accepted in final form March 7, 2022. Submitted and externally peer reviewed. The handling editor was Friedemann Paul, MD.

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## Appendix (continued)

Name	Location	Contribution
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Zélia Poulet, Julie Pique, Adil Maarouf, et al.  
*Neurol Neuroimmunol Neuroinflamm* 2022;9;  
DOI 10.1212/NXI.0000000000001167

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