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## Cranial mononeuritis multiplex as the initial manifestation of systemic lupus erythematosus: A diagnostic challenge<sup>☆</sup>



### Multineuritis craneal como comienzo de lupus eritematoso sistémico: un reto diagnóstico

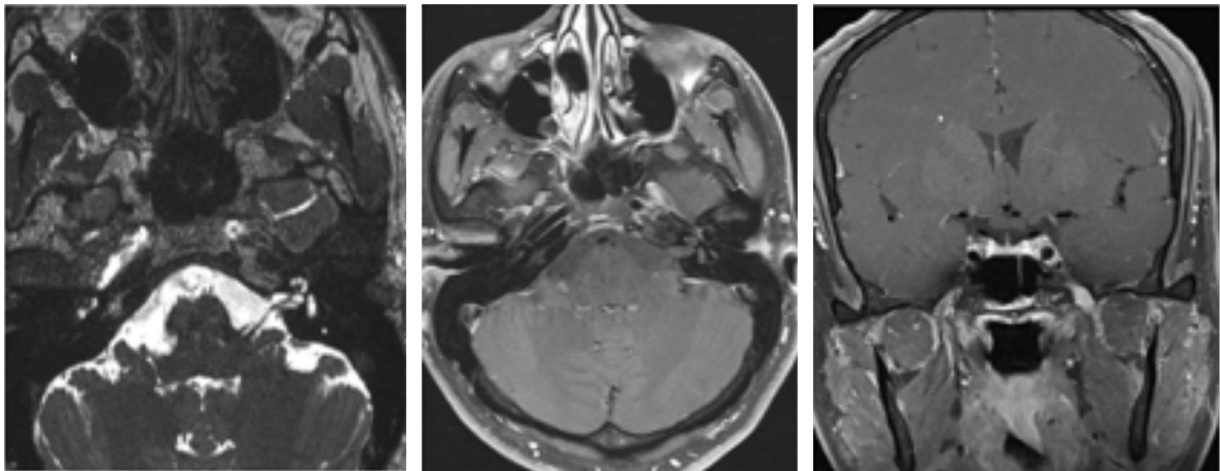
*Dear Editor:*

Mononeuritis multiplex (MM) is an infrequent disorder that may be secondary to a number of diseases, which makes aetiological diagnosis difficult.<sup>1</sup> Infectious and inflammatory aetiologies may be difficult to differentiate. Magnetic resonance imaging (MRI) and cerebrospinal fluid (CSF) analysis are useful tools for determining the cause of MM. We present the challenging case of a patient with MM whose symptoms were initially thought to be associated with herpes simplex virus (HSV) infection but who later developed the typical symptoms of systemic lupus erythematosus (SLE). Very few cases of MM secondary to SLE have been described in the literature<sup>2</sup>; cases of MM as an initial manifestation of SLE are even rarer. Our patient was a 42-year-old man with a history of hemifacial spasm due to left-sided facial paralysis in adolescence and recurrent symptoms of oral herpes virus infection.

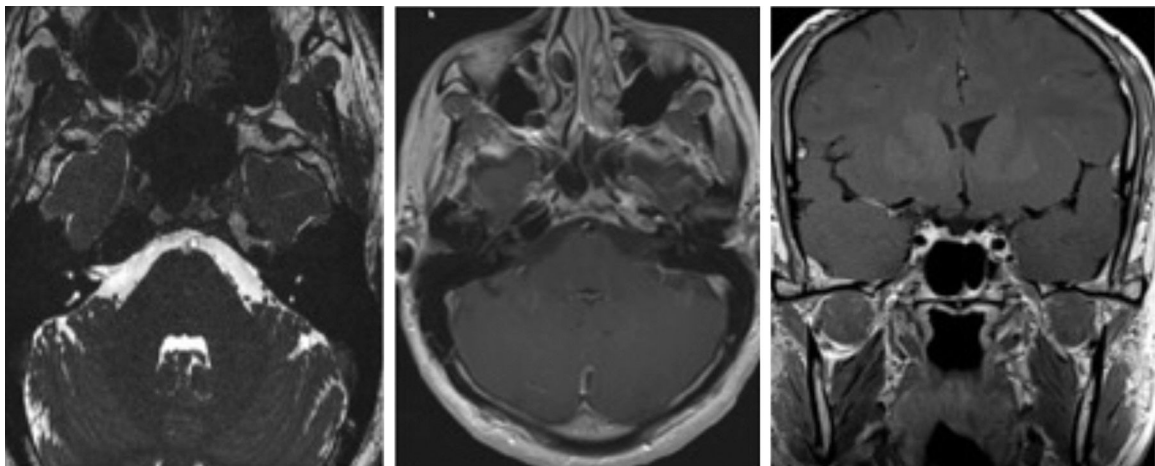
The patient came to our department due to left-sided facial dysaesthesia, which was interpreted as neuralgia of the second branch of the left trigeminal nerve. Symptomatic treatment achieved little improvement. A brain MRI scan revealed thickening of the left trigeminal nerve (Fig. 1). Our patient had experienced recurrent symptoms of oral herpes virus infection a month previously; the MRI findings were therefore thought to be due to HSV reactivation. The patient returned to our department 9 months later, reporting worsening of the dysaesthesia; he also presented diplopia, secondary to left sixth cranial nerve palsy. We performed an additional brain MRI scan: thickening of the second branch of the trigeminal nerve persisted, with the sixth cranial nerve showing no alterations. A CSF analysis disclosed no relevant findings,

viral serology results were negative, and the results of an immunological study were negative for antiphospholipid antibodies and positive for antinuclear antibodies (1:320). The patient was not receiving any drugs that may have induced lupus erythematosus. Due to suspicion of MM of inflammatory origin, we started treatment with megadose methylprednisolone therapy, achieving symptom improvement after 3 weeks of treatment. The patient was examined by the rheumatology department, reporting a 6-month history of arthritis affecting the elbows and lower limbs, which prevented him from exercising, a 2-month history of mouth ulcers, and photosensitivity. These symptoms, together with the presence of positive antinuclear antibodies, pointed to SLE.<sup>3,4</sup> The patient was diagnosed with SLE as the initial manifestation of MM, and started treatment with cyclophosphamide; symptoms disappeared and the MRI findings of nerve thickening and contrast uptake resolved (Fig. 2). Around 50% of patients with SLE display central nervous system (CNS) involvement.<sup>5</sup> This usually occurs 2 years after diagnosis of SLE.<sup>4</sup> Cranial nerve palsy is infrequent, accounting for 0.5%-1% of all neuropsychiatric manifestations.<sup>5</sup> Most of the cases published in the literature report isolated cranial nerve palsies, with cranial MM being extremely rare.<sup>2</sup> Cranial nerve palsy is rarely associated with SLE; palsies are normally secondary to a concomitant central or peripheral nervous system disorder (meningitis, Guillain-Barré syndrome, or cavernous sinus inflammation). In many cases, the pathogenic mechanism of cranial neuropathies in SLE is difficult to determine. Several mechanisms have been proposed; the most widely accepted is antibody-mediated neuronal involvement, which causes vasculopathy, intrathecal production of proinflammatory cytokines, and accelerated atherosclerosis.<sup>6</sup> It is uncertain whether the pathogenic mechanism of SLE is inflammatory, thrombotic, or mixed. The eighth and tenth cranial nerves are the most frequently affected in SLE.<sup>7</sup> The literature describes 2 cases of isolated twelfth cranial nerve involvement secondary to SLE<sup>8,9</sup> and 3 cases of isolated fifth cranial nerve involvement.<sup>10</sup> Optic neuropathy is the most frequent neuro-ophthalmological manifestation of SLE.<sup>7</sup> Ocular involvement secondary to cranial nerve palsy is rare, with the sixth cranial nerve being the most frequently involved.<sup>7</sup> Regarding MM, Keane<sup>2</sup> examined the causes of multiple cranial neuropathy in 979 patients and found only one case secondary to SLE. The literature includes 2 cases of MM associated with SLE: one patient with SLE and sarcoidosis who displayed third and sixth cranial nerve involvement combined with peripheral neuropathy,<sup>5</sup> and another patient with SLE with neuro-ophthalmological

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**Figure 1** Axial T2-weighted and axial postcontrast fat-suppressed T1-weighted MR images display uniform thickening and contrast uptake in the maxillary branch of the left trigeminal nerve from the Meckel cave to the pterygopalatine fossa. Coronal T1-weighted sequence showing nodular thickening of the mandibular branch of the left trigeminal nerve at the foramen ovale.



**Figure 2** A follow-up MRI scan performed at 4 months revealed resolution of contrast uptake and thickening of the mandibular and maxillary branches of the trigeminal nerve.

manifestations, who was diagnosed with left-sided MM secondary to SLE.<sup>7</sup> To our knowledge, this is the first case of MM associated with fifth and sixth cranial nerve involvement as the initial manifestation of SLE. Immunosuppressive treatment (cyclophosphamide or azathioprine) and corticosteroids are indicated for inflammatory MM. Patients with positive antiphospholipid antibodies should also receive anticoagulation therapy.<sup>6</sup> Our patient responded well to cyclophosphamide. No diagnostic algorithm is available for MM due to the lack of evidence-based guidelines. In these cases, we should aim to detect biomarkers or neuroimaging findings that might identify the underlying pathological mechanism. SLE should be considered in the differential diagnosis of MM.

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### Conflicts of interest

The authors have no conflicts of interest to declare.

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## Cognitive profile of a child with SOS1 mutation in Noonan syndrome<sup>☆</sup>



## Perfil cognitivo de un niño con síndrome de Noonan con mutación genética SOS1

*Dear Editor:*

We present the neuropsychological profile of a 7-year-old boy with Noonan syndrome (NS) and *SOS1* mutation.<sup>1</sup> Studies on NS usually present a general, non-specific image of cognitive impairment; in our opinion, this is due to the limited number of studies and the small samples included, which do not accurately represent the variability of the condition, since they group together patients with different mutations.<sup>2–5</sup> The case we present is interesting due to the scarcity of literature on the differential cognitive characteristics observed in this mutation, in comparison with the disability reported in other, more frequent mutations.

We present the case of a boy who was diagnosed with NS at the age of 4 by means of the polymerase chain reaction method. The medical history suggested normal development of motor, communication, and language skills.

We assessed his performance in several cognitive processes. His general ability index was 131, and composite score (CS) was 98 on the Wechsler Intelligence Scale for Children (WISC-IV).

**Attention.** The patient obtained normal results in information processing speed (Conners' Continuous Performance Test II [CPT-II] version 5: CS = 61) and in cue-based tasks in

WISC-IV (scaled score [SS] = 11). He scored poorly (SS = 6) in the symbol search subtest, which indicates difficulties in visual perception, confirmed by scores below the mean on the Illinois Test of Psycholinguistic Abilities (ITPA) (*T*-score = 26). Variability of response time to stimuli (CPT-II: SS = 51) and attentional vigilance (CS = 50) were normal. Results for selective and focused attention (CPT-II) were normal, but with many omissions (CS = 99), suggesting inattention. Results for response style were average (CPT-II: CS = 55).

**Perception.** The patient obtained average results for visual discrimination (CPT-II score = 51). In the perceptual reasoning subtests (WISC-IV), he scored highly in block design (SS = 14) and picture concepts (SS = 12), and very highly in matrix reasoning (SS = 19). Visual processing (WISC-IV) scores were average in the block design + picture completion tests (CS = 91). In the ITPA, he obtained lower scores on the visuomotor channel than on the auditory-vocal channel. Concept visual comprehension was very good (*T*-score = 43, *M* = 35) and comprehension of relationships between visual stimuli was average (*T*-score = 34). At the automatic level, he scored poorly for visual integration (*T*-score = 26).

**Memory.** Regarding working memory (WISC-IV), he obtained low-normal scores on the digit span (SS = 8). He obtained a slightly below-average score for visual sequential memory (ITPA: *T*-score = 32). Results in WISC-IV subtests for long-term memory (CS = 99.6) and general information (CS = 96) were excellent.

**Language.** Scores for verbal comprehension in the WISC-IV were high in similarities (SS = 14), very high in vocabulary (SS = 19), and average in the comprehension subtest (SS = 11). In fluid verbal reasoning, he obtained high average scores (CS = 95). Scores for lexical knowledge were high (99.6). The results on the auditory-vocal subtests in the ITPA representational level indicate an age equivalent to his age (mean = 35). Story retelling (*T*-score = 36, *M* = 35) and verbal analogies (*T*-score = 36) results were normal. He obtained average scores (STEN score = 6) in verbal expression in the ENFEN test battery. In the automatic level of the ITPA, the

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