

Analysis of central nervous system findings on MR imaging in patients with neurolupus

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Purpose

To study the frequency and the vascular cerebral pattern of central nervous system involvement in patients with neurolupus.

Background.

#The **prevalence of systemic lupus erythematosus (SLE)** is up to 150/100.000, depending principally on the gender and race (5-10 times more frequent in women and in Afro-Caribbean and Asian population) [1, 2].

#The **diagnostic of SLE** is based on the American College of Rheumatology (ACR) diagnostic criteria, requiring a minimum of 4 out of 11 [1].

#**Neurolupus** has a **prevalence** of 14-90% of all the SLE patients during the entire course of their disease, with a generally accepted average of 30-40%. Also it represents the **cause of death** in 19% of SLE patients [1, 2].

#According to the ACR criteria, **neurolupus includes 12 syndromes**, divided in *neurological* (aseptic meningitis, cerebrovascular lesions, demyelinating syndromes, headache, movement disorders, myelopathy, epilepsy) and *neuropsychiatric* (acute confusional states, anxiety disorder, cognitive dysfunction, affective disorders) [2].

#

The average age of the patients with **cerebral infarct due to SLE** is 35 years, with a recurrence in 35-60% of the patients associated to the presence of antiphospholipidic antibodies [1, 2].

#One of the most debilitating complications of neurolupus is the **myelopathy**. It has a prevalence of 1-5% of the SLE patients. It usually develops early in the evolution of the disease leading to an unfavorable prognosis. In 39% of the patients with lupus related myelopathy it constitutes the presenting symptom of SLE, and in another 42% occurs during the first 5 years after the diagnostic [2, 3].

Pathology of cerebral lesions in SLE:

- *microangiopathic disease* (the most frequent neuropathological finding; due to intimal hyperplasia, erythrocytes extravasation and development of fibrin thrombi). It typically produce a multifocal involvement, sometimes with multiples microinfarcts
- *macroscopic infarcts* (less frequently, principally explained by secondary coagulopathy due to antiphospholipidic antibodies or by embolic phenomena due to Libman-Sacks endocarditis)
- *accelerated atherosclerosis* (steroid treatment, vasculitis and microhemorrhages playing a role)
- *direct autoimmune neural alteration, demyelination, embolisms [1-3].*

In the medical literature there is a lack of studies with large number of neurolupus patients describing the characteristics and prevalence of brain lesions in this population.

The two **most frequent MR findings** reported in **SLE patients** were: **multiple small lesions** (8-70%) and **brain atrophy signs** (9-67%). They are commonly found in patients with long course disease and in those with neurolupus [2, 3].

Methods and Materials

Retrospective descriptive study. We carried out a review of 40 patients with neurolupus, studied by MRI during the last 7 years (2006-2012).

The MRI studies were performed using three different systems of 1,5 T and 3 T and included T1, T2, FLAIR sequences and in 10 patients also diffusion sequences.

The lesions were classified according to:

- Distribution and anatomical location:

-Supratentorial:

1. # periventricular white matter
2. # deep white matter
3. # corpus callosum
4. # cortico-subcortical (great territory infarcts)

#-Infratentorial:

1. spinal cord
 2. brainstem
 3. cerebellum
- #Number: unique, double and multiple
 - #Size: less than 1 cm, between 1 and 5 cm, more than 5 cm.

We used the Fazekas scale to determine the degree of white matter involvement (examples of Fazekas grades in [Fig. 1](#) on page 5).

We performed a descriptive statistical study using SPSS 15.0.

Images for this section:

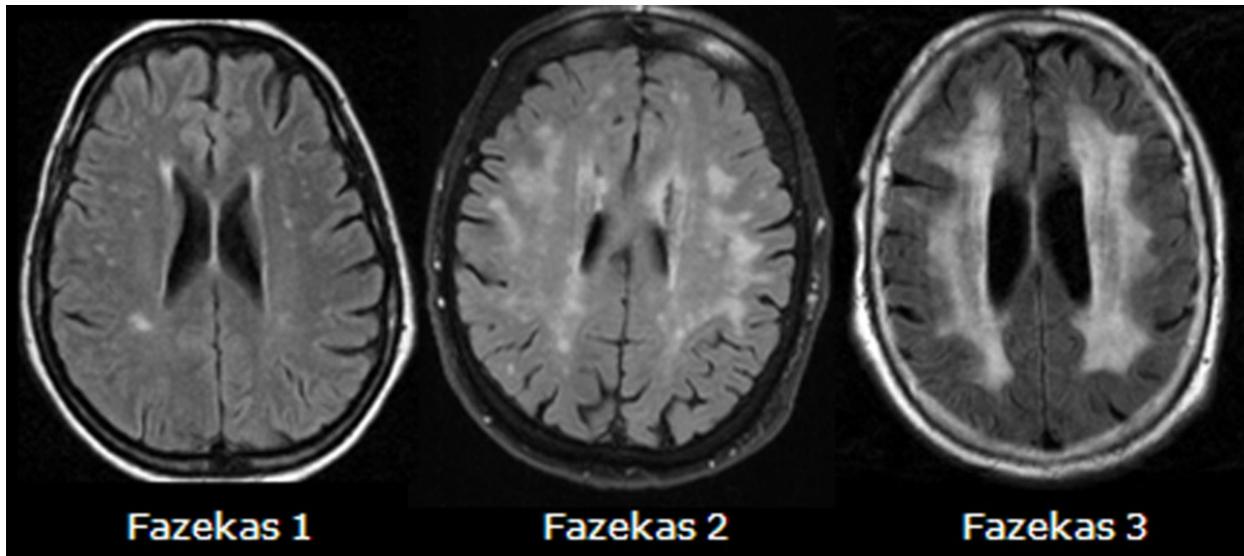


Fig. 1: Fazekas grading scale of white matter lesions (examples)

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Results

The average age was 43 years (range 18 to 83). Thirty-seven (92,5%) were women and 3 (7,5%) men.

We found cerebral anomalies in 14 patients (35%). In this group the mean age was 43,7 years (range 27 to 66).

Of the 14 patients who had MRI findings, 9 (64,3%) presented with only supratentorial involvement, 4 (28,6%) had both supra and infratentorial lesions and 1 only infratentorial.

#Of the 5 patients with infratentorial involvement, the location of the lesions was: cerebellum in 3 patients, brainstem in 3 and spinal cord in 2.

The supratentorial lesions were located in the deep white matter in 11 (78,5%), the periventricular area in 7 (50%), the corpus callosum in 2 (14,2%) and in cortico-subcortical areas in 2 (14,2%) patients (Fig. 2 on page 8).

Related to Fazekas scale, 12 (85,7%) were graded as 1 and 2 (14,3%) graded as 2. There were no grade 3 patients.

Twelve patients (85,7%) presented multiple lesions and 2 (14,3%) had single lesion.

In 10 patients (71,4%) the lesion size was less than 1 cm and in 4 (28,6%) between 1 and 5 cm. Two (14,3%) patients had extensive lesions (more than 5 cm), both of them in posterior fossa (Fig. 3 on page 8).

Examples of neurolupus cases:

Case 1 - 39 year-old woman, diagnosed with neurolupus, with small periventricular white matter lesions and previous right parietal infarct (Fig. 4 on page 9).

Case 2 - 66 year-old woman with neurolupus

- right subcortical parietal and deep temporal (hippocampus) white matter lesions, without contrast-enhancement, without diffusion restriction, of inflammatory aspect (vasculitis/encephalitis)

- periventricular lesions and of the splenium of the corpus callosum

- vasogenic edema of the basal ganglia with infarct with diffusion restriction of the right lenticular
- involvement of the right part of the pons ([Fig. 5](#) on page 10).

Case 3 - 51 year-old woman, diagnosed with neurolupus, with periventricular lesions simulating the typical aspect of multiple sclerosis ([Fig. 6](#) on page 11).

Case 4 - 27 year-old woman with neurolupus and extensive myelitis (cervical, thoracic and bulbar level), without supratentorial involvement ([Fig. 7](#) on page 11).

Images for this section:

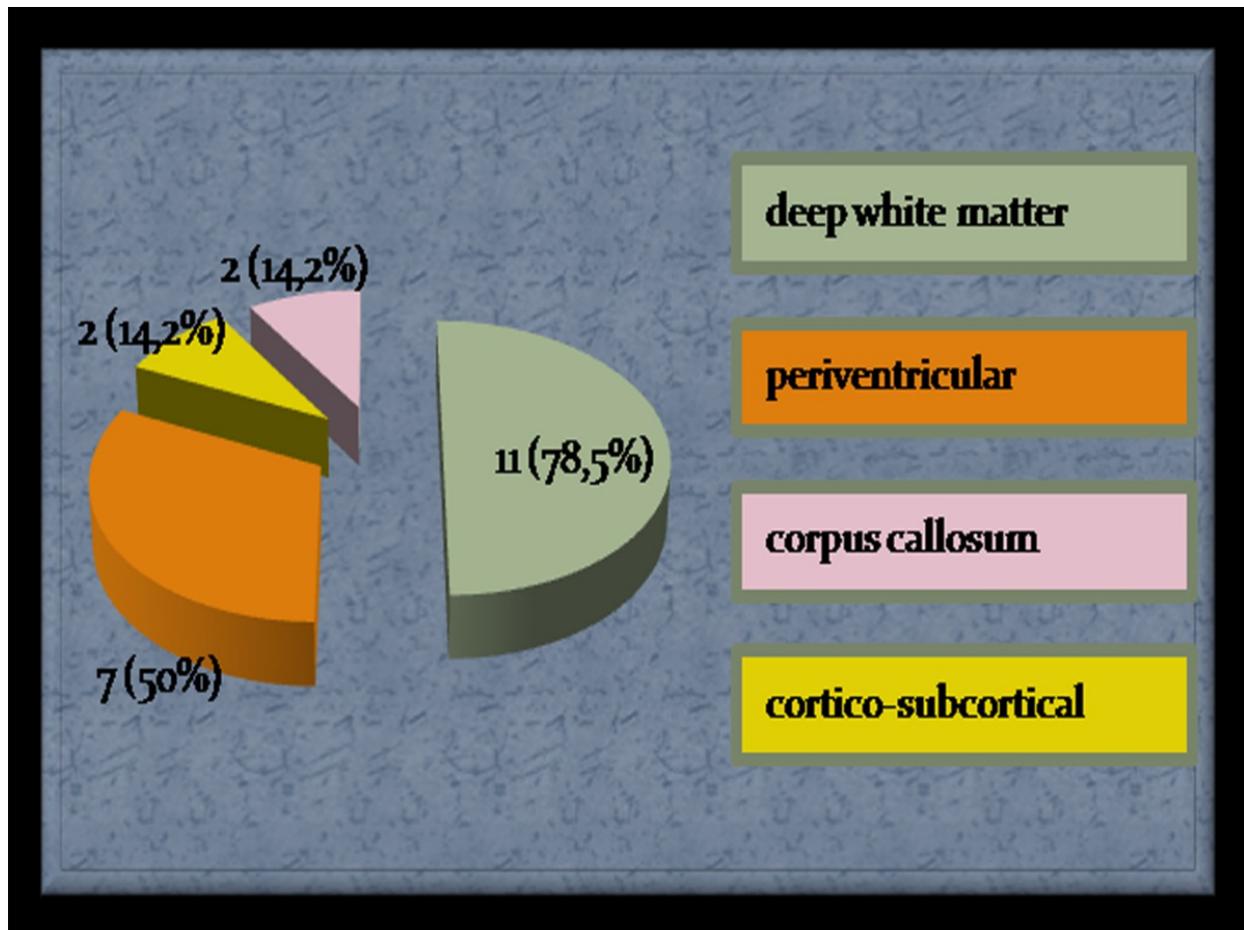


Fig. 2: Anatomical location of the supratentorial lesions

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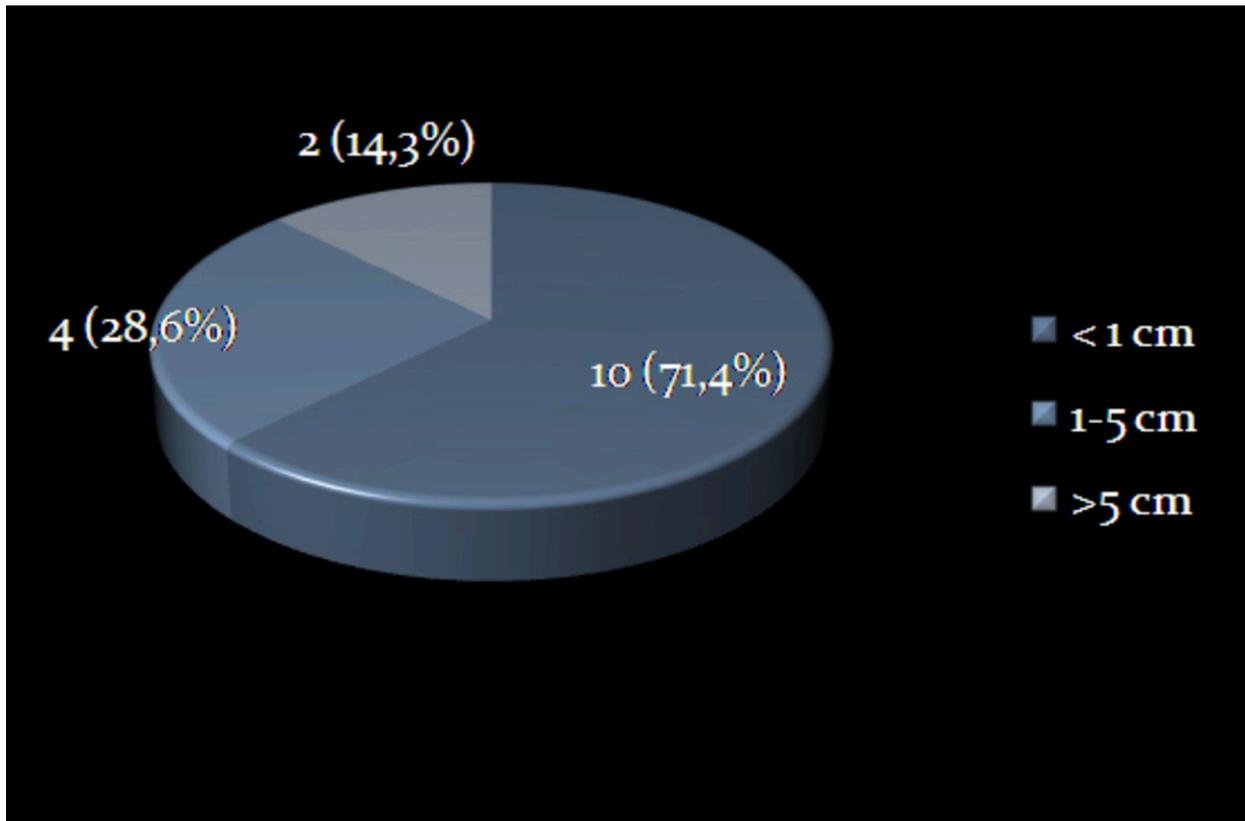


Fig. 3: Lesion size

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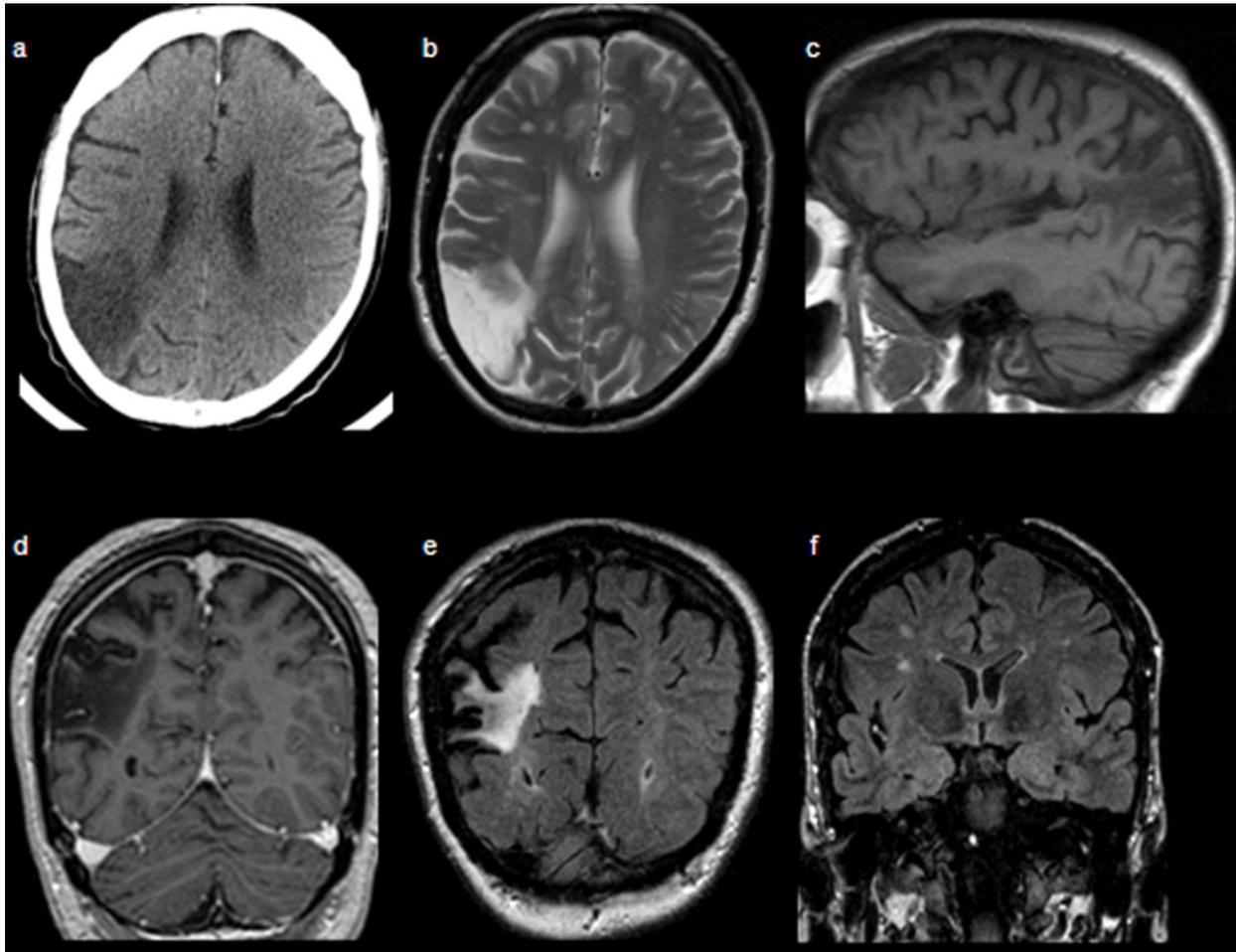


Fig. 4: Axial nonenhanced CT (a), axial T2WI (b), sagittal T1WI (c), coronal contrast-enhanced T1WI (d) and coronal FLAIR (e, f) showing small periventricular white matter lesions, Fazekas grade 1, and right parietal encephalomalacy corresponding to a previous infarct in the posterior superficial territory of the right middle cerebral artery

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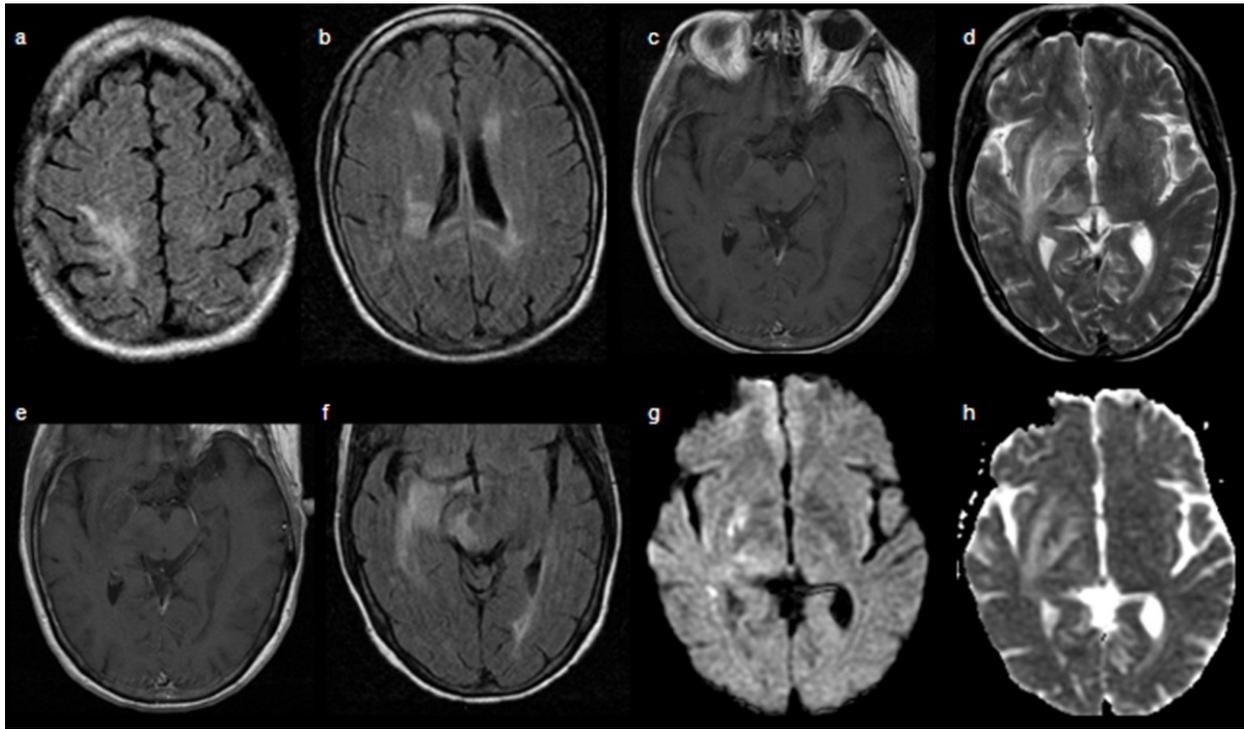


Fig. 5: Axial FLAIR (a, b, f), contrast-enhanced T1WI (c, e), T2WI (d) and DWI/ADC maps (g, h) showing: extensive lesions of the right parietal and temporal white matter, without contrast-enhancement, without diffusion restriction, of inflammatory aspect (vasculitis/encephalitis); periventricular lesions and of the splenium of the corpus callosum; vasogenic edema of the basal ganglia and infarct with diffusion restriction of the right lenticular; involvement of the right part of the pons

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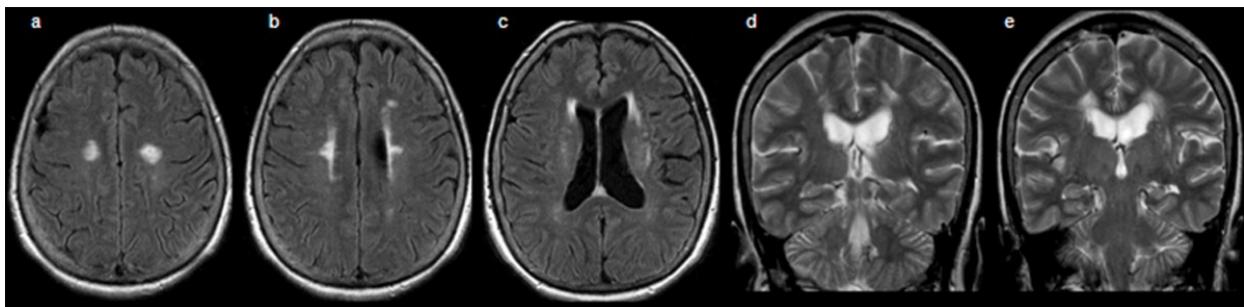


Fig. 6: Axial FLAIR (a-c) and coronal T2WI (d, e). Periventricular beginning confluent lesions (Fazekas grade 2) simulating the typical aspect of multiple sclerosis

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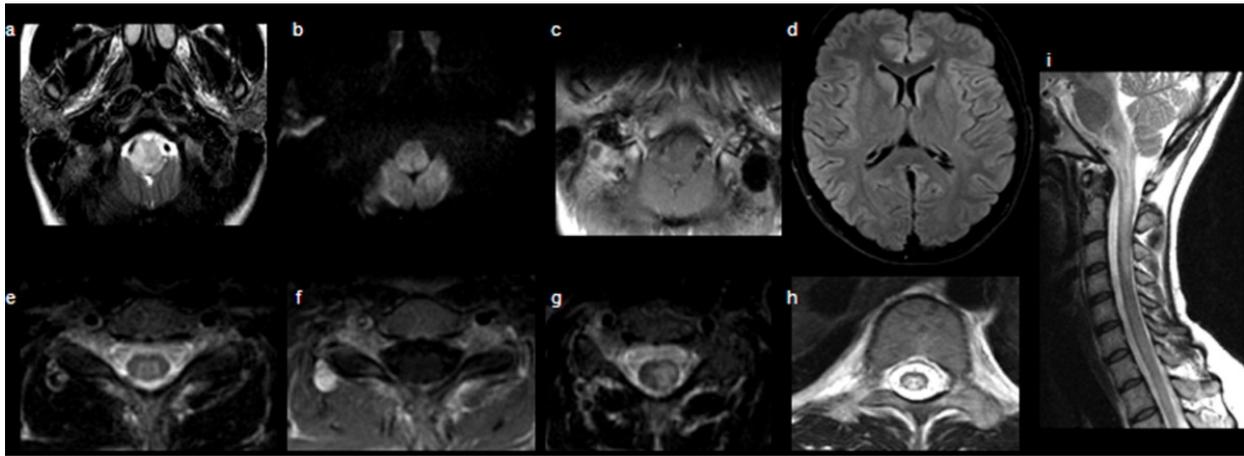


Fig. 7: Extensive myelitis involving the medulla (axial T2WI, FLAIR and T1WI in a-c), cervical spine (axial T2WI in e, g and T1WI in f) and thoracic segment (axial T2WI in h). Sagittal T2WI (i) showing a better depiction of the extension. Axial FLAIR at supratentorial level (d) showed no lesions

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Conclusion

A non depreciative number of neurolupus patients have MRI findings, the most frequent pattern being small supratentorial white matter lesions, representing small vessel disease.

References

1. Joseph FG, Scolding NJ. Neurolupus. Pract Neurol. 2010;10:4-15.
2. Katsumata Y, Harigai M, Kawaguchi Y, et al. Diagnostic reliability of magnetic resonance imaging for central nervous system syndromes in systemic lupus erythematosus: a prospective cohort study. BMC Musculoskeletal Disorders. 2010;11-13.
3. Appenzeller S, Pike GB, Clarke AE. Magnetic resonance imaging in the evaluation of central nervous system manifestations in systemic lupus erythematosus. Clin Rev Allergy Immunol. 2008;34:361-6.

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