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Dropped head syndrome in systemic lupus eritematoso (SLE): Case report and review of the neurological manifestations of LES

Oliveira FTM[1]*, Pellegrinelli A[1] and Gennari JD[2]

[1]Department of Neurology, Santa Casa of São Paulo-SP, Brazil

[2]Department of Rheumatology, Santa Casa of São Paulo-SP, Brazil

Abstract

The dropped head syndrome (DHS) is a weakness of neck extension, leading to a typical presentation of the flexed head with the chin in contact with the chest wall. We report a case of a patient with DHS due to vasculitis of the CNS secondary to SLE and performed a brief review of the literature on the subject. SLE is a systemic disease with several neurological manifestations, the prompt recognition of these conditions usually leads to a better long-term prognosis from a functional point of view. We show the main complications and an example of a successful case due to the prompt care of the neurology and rheumatology.

Case report

L.B.O., 38 years old, in follow-up at our service by SLE, with involvement of CNS previously (cerebral vasculitis secondary to lupus activity), using corticoid therapy and immunosuppressant (azathioprine). Patient in clinical stability and in weaning of corticoid, presented with clinical picture of fallen head, dysphagia, dysphonia and tetraparesis, with muscular strength global IV, deep tendon reflexes 4+ global, and superficial hypoesthesia, with a sensitive level in C6. He underwent partial-response methylprednisolone pulse therapy, evolving with septic pulmonary focus. After intravenous cyclophosphamide treatment, the patient presented motor and sensory improvement. The study of cerebrospinal fluid by lumbar puncture showed inflammatory activity, without changes in the research of infectious agents. Performed MRI of the brain and cervical and thoracic columns, which showed signs suggestive of cerebral vasculitis and cervical spinal cord.

Introduction

Systemic Lupus Erythematosus (SLE) is a chronic systemic autoimmune disease of connective tissue with various clinical manifestations, characterized by periods of exacerbation and remission. This disease has a marked characteristic, the exacerbated production of antibodies (anti-DNA, anti-Sm, anti-RNP, anti-Ro / SSa, anti-La / SSb, among others).

There are no definitive criteria for the diagnosis of SLE. The American College of Rheumatology defined classification criteria for SLE, according to which at least four clinical and/or laboratory criteria are necessary among eleven after careful investigation and exclusion of infectious, neoplastic, and other diseases [1,2].

The criteria consider the following manifestations:

- Malar rush
- Discoid lesion
- Photosensitivity
- Oral mucous ulcers
- Non-deforming arthritis
- Serositis (pleuritis, pericarditis).
- Renal disease (persistent proteinuria, cylindrury).
- Involvement of the central nervous system (convulsion and psychosis).
- Hematological changes (anemia, leucopenia or thrombocytopenia).
- Immunological changes: anti-DNA, anti-Sm or false positive VDRL.
- Anti-nuclear factor (ANF)

Neurological manifestations of SLE

Neurological manifestations in SLE are complex and can be defined as: neurological manifestations of the central nervous system (CNS), peripheral (PNS), autonomic (ANS) and psychiatric syndromes.

These manifestations may be caused directly by SLE activity or be secondary to comorbidities such as systemic arterial hypertension (SAH), diabetes mellitus (DM), uremia, and infection.

Patients with SLE may present CNS involvement with a very variable frequency, 14 to 75%, depending on the population studied and the inclusion criteria used. Probably the main reason for the huge statistical differences found in the frequency of CNS involvement is the inclusion of minor non-specific symptoms.

Due to the fact that clinical manifestations may be caused directly by SLE activity or occur secondary to comorbidities, it is difficult to

Correspondence to: Oliveira FTM, Department of Neurology, Santa Casa at São Paulo- SP, Av. Dr Cesario Mota Jr, 112 – V. Buarque. São Paulo-SP, Brazil, CEP 01221-010; E-mail: towmaz@gmail.com

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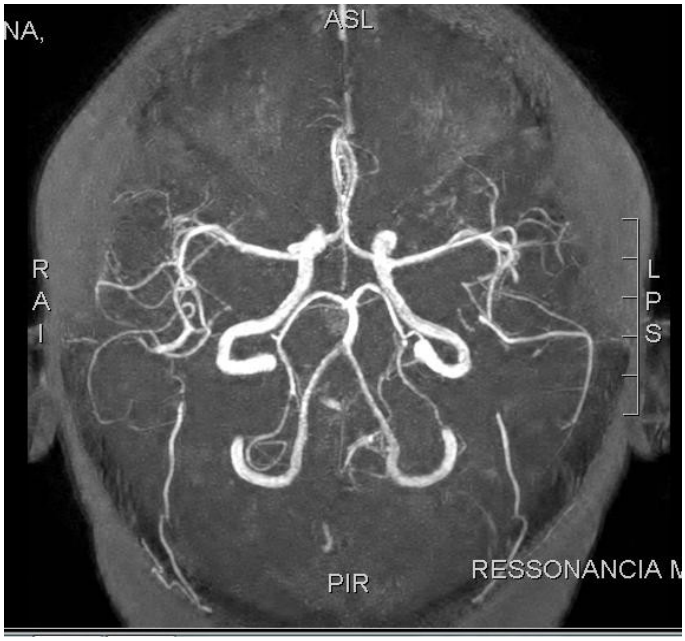


Figure 1. Angiographic study of venous drainage of the encephalon without alterations and intracranial arteries with inflammatory vasculopathy.

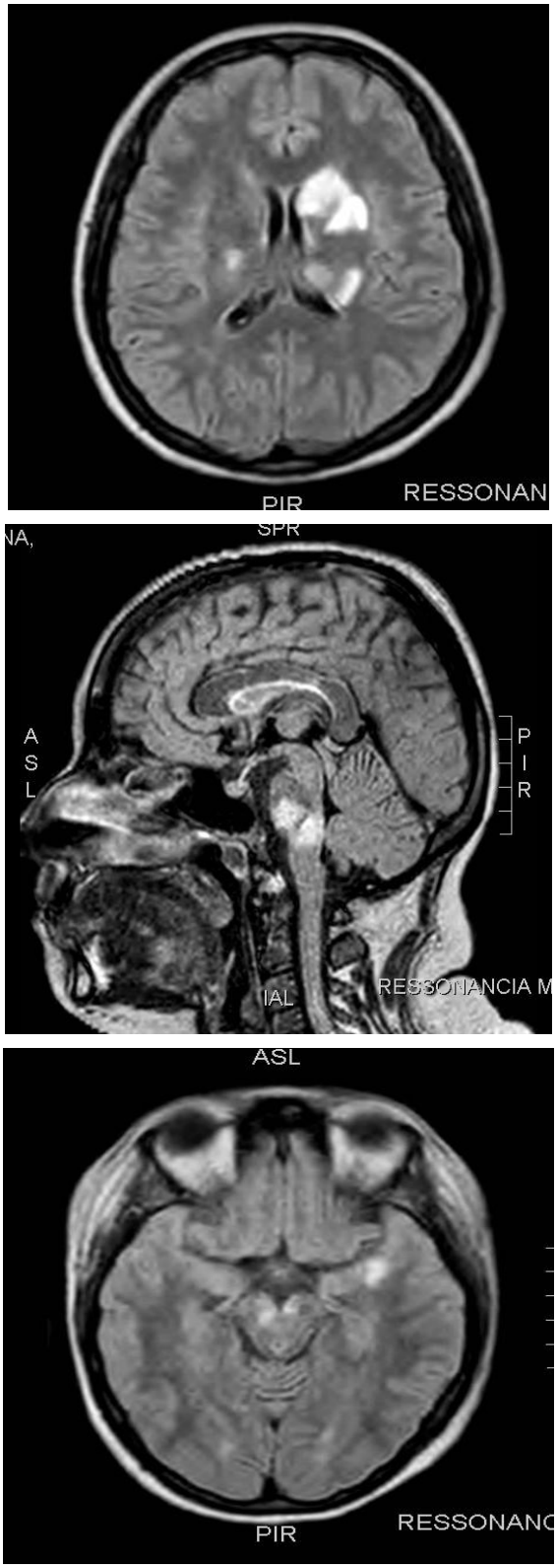
define which changes should be attributed directly to SLE, or which are secondary to the treatment performed or to other associated conditions.

The neuropsychiatric manifestations of SLE are divided into 2 subgroups, according to the American College of Rheumatology (ACR) in 1999: CNS manifestations (headache, convulsion, anxiety disorder, mood disorder, movement disorders, cognitive disorders, cerebrovascular disease, confusional state acute, aseptic meningitis, myelopathy, psychosis, demyelinating syndromes) and manifestations of PNS (autonomic disorder, Myasthenia Gravis, mononeuropathy, cranial neuropathy, plexopathy, polyneuropathy, acute demyelinating inflammatory polyradiculopathy). Major CNS manifestations according to RTA definitions would include: chorea, aseptic meningitis, psychosis, convulsions, myelopathy, demyelinating syndrome, acute confusional state, and stroke [1-9].

The importance of early recognition, clinical and laboratorial analysis and adequate treatment of CNS manifestations in SLE are due to the influence of this influence on mortality, quality of life and permanent functional impairment.

The difference in the numbers of CNS involvement in the literature shows that a better study of this involvement is important, mainly because the long-term repercussions of this possibly underdiagnosed condition can lead to a great functional compromise and the quality of life of the CNS. patients. This involvement of the CNS in SLE is complex because it involves multiple clinical presentations. The ACR classified these changes into 19 syndromes, 12 of which were CNS (major: aseptic meningitis, cerebrovascular diseases, demyelinating syndrome, movement disorders, epilepsy, myelopathy, psychosis, acute confusional state, and minor ones: headache, anxiety disorder, mood and cognitive impairment) and 7 of the PNS (Guillain Barré syndrome, autonomic neuropathy, mononeuropathy, myasthenia gravis, cranial neuropathy, plexopathy, polyneuropathy).

Histopathological studies in patients with SLE have often found a vasculopathy of small intracranial vessels consisting of proliferative changes of the intima, vascular hyalinization and perivascular



Figures 2 to 4. Multiple signal alteration foci characterized by the hypersignal in T2 and FLAIR located in the trunk of the corpus callosum, nucleocapsular regions, thalamus. The parenchymal changes observed in the thalamus and posterior arm of the internal capsule (nodular enhancement to paramagnetic contrast medium and restriction to the free movement of water molecules, compatible with foci of subacute ischemia) should result from probable inflammatory vascular changes related to the underlying disease inflammatory vasculopathy.

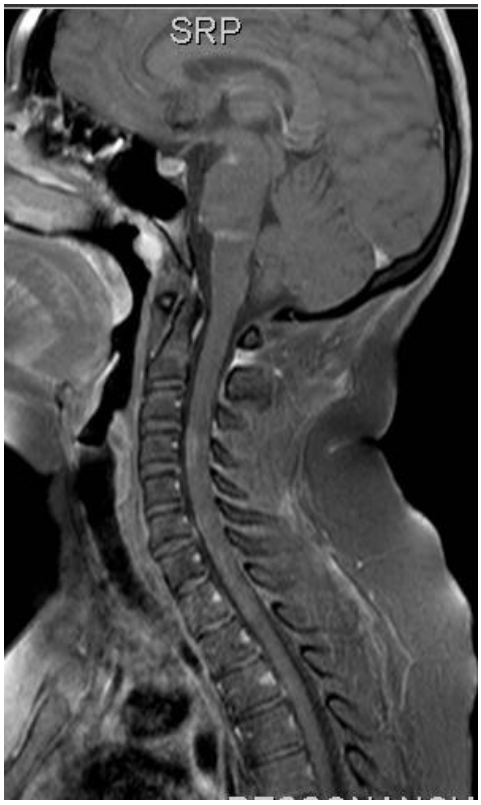


Figure 5. Signal alteration compromising the spinal cord at C3-C4 levels in T2 / FLAIR, with impregnation by the paramagnetic contrast agent, which should result from inflammatory vascular changes related to the underlying disease, corroborating the hypothesis of inflammatory vasculopathy (vasculitis).

lymphocytosis. This vasculopathy of small vessels was seen in both SLE with psychiatric symptoms and in those with focal manifestations. Studies with SPECT and MRI of the skull with spectroscopy suggest that both cerebral atrophy and cognitive decline in SLE patients may be related to chronic cerebral schistosomiasis [10-12].

The production of autoantibodies has been implicated in this vasculopathic process and associated with neuronal injury, mainly anti-P antibodies and antiphospholipid antibodies, with a strong correlation with the major manifestations of CNS involvement in SLE [12-14].

Antiphospholipid antibodies are members of a family of antibodies directed against plasma proteins bound to negatively charged phospholipids, which lead to hypercoagulability through effects on protein C, protein S, platelets, endothelial cells and complement activation. Antiphospholipid antibodies, defined as lupus anticoagulant, anticardiolipin and anti-b2 glycoprotein-1 (anti-b2GP1), are strongly associated with transient ischemic attack (TIA), stroke, seizures, and cerebral venous thrombosis [15].

Cerebral ischemic events may occur early in the course of SLE, or may even precede the diagnosis, leading to the diagnosis of underlying SLE. The frequency of cerebrovascular disease as a whole ranges from 5.3 to 19%, depending on the study methodology and the neuropsychiatric syndromes included in the series. Cerebral venous thrombosis is a rare complication, and as well as hemorrhagic stroke, intracerebral hemorrhage and arachnoid, can also occur in SLE. The association between Antiphospholipid Syndrome and venous or arterial thrombotic events has been described in both the primary antiphospholipid syndrome and the secondary SLE [16-20].

Cranial neuropathies occur more frequently from II to VII cranial nerves, causing ocular manifestations, either as isolated internuclear ophthalmoplegia or as paralysis of II, IV or VI nerves.

A demyelinating syndrome can cause motor deficit in one or more limbs, secondary to transverse myelopathy, or diplopia due to optic neuropathy, isolated nerve palsy or internuclear ophthalmoplegia.

Some studies are recommended in the evaluation of all patients with possible demyelinating syndrome, including evaluation:

1) CSF: for cell counts, proteins, oligoclonal bands, IgG index, cultures and cytology;

2) MRI of the brain, and according to the clinical picture, evoked potentials.

The differential diagnosis of demyelinating syndromes includes structural lesions, familial disorders such as ataxia and leukodystrophies, sarcoidosis, Behçet's syndrome, optic neuromyelitis and its spectrum of presentations, multiple sclerosis, and the exclusion of infectious diseases (tuberculosis, HIV, syphilis) and deficits nutritional deficiencies (vitamin B12 deficiency).

Probably the main reason for the huge statistical differences found in the frequency of CNS involvement is the inclusion of minor non-specific symptoms, including headache, mild cognitive impairment and depressive symptoms, which may occur in several chronic autoimmune diseases, patients treated with steroids and immunosuppressants, with difficult characterization as disease activity, independent diseases or consequence of the various associated comorbidities. Hence the importance of larger studies showing the actual frequency of major manifestations of CNS involvement by SLE [13,21-30].

The presentation of DHS in a patient with SLE is relatively rare, being more common in patients with neurological diseases, such as [31]:

- Degenerative diseases: Amyotrophic lateral sclerosis, Parkinson's disease, Multiple system atrophy;

- Movement disorders: cervical dystonia, tardive dyskinesia;

- Neuromuscular: Myasthenia Gravis, Post-polio syndrome, Lambert-Eaton myasthenic syndrome, Chronic inflammatory polyradiculoneuropathy. Muscle involvement may be inflammatory or non-inflammatory, or secondary.

- Inflammatory primary muscle: polymyositis;

- Non-inflammatory primary muscle: nemaline myopathy, inclusion corpuscle myopathy, mitochondrial myopathies, congenital myopathies;

- Secondary muscle: post-radiation, post-toxin botulinum, hypokalemia, hypothyroidism.

Conclusion

We present a rare case of DHS secondary to CNS involvement due to cervical myelitis secondary to vasculitic activity. We verified through this broad review that the SCC has several etiologies (myogenic, neurogenic, metabolic, degenerative and local causes), and that within the context of rheumatologic autoimmune diseases, an extensive investigation becomes important because the therapeutic behavior can vary widely, including broadly altering the long-term functional prognosis of these patients.

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