Case study

Longitudinal transverse myelitis with locked in syndrome revealing a systemic lupus erythematosus

Abstract :

The neuropsychiatric manifestations of lupus are very heterogeneous and are a source of significant morbidity and mortality. Transverse myelitis is a rare but serious complication of systemic lupus erythematosus, classically described as an acute attack with a poor functional prognosis. It most often complicates the course of a previously diagnosed lupus and corresponds clinically to a complete medullary syndrome associating a symmetrical bilateral sensitivo motor deficit and sphincter disorders. We report a rare case of a young woman who was admitted to intensive care for a rapidly evolving flaccid tetraparesis with impaired consciousness. The explorations concluded that there was lupus with an inaugural neurological disorder such as longitudinal transverse myelitis associated with a secondary locked-in syndrome and a pontine ischemic stroke. Our observation has several particularities: the inaugural character and the longitudinal form of lupus myelitis as well as the serious association at the outset with a locked-in syndrome secondary to a cerebrovascular accident.

Key words : Systemic lupus erythematosus, myelitis, neurology.

Introduction :

Systemic lupus erythematosus (SLE) is a multi-system autoimmune disease of connective tissue. There is a wide variety of neurological signs, from mild visual disturbances to severe central nervous system damage. Cerebrovascular disease, mainly ischemic strokes, occurs in almost 10% of lupus patients (1). Transverse myelitis is rare in SLE, reported in 1% to 2% of cases (2). It usually occurs after years of development and is exceptionally indicative of the disease. Longitudinal myelitis is a rare form of transverse myelitis, characterized by a serious clinical symptoms and poor prognosis.

Aims :

We report a serious acute case of longitudinal transverse myelitis associated with a locked-in syndrome (LIS) secondary to a pontine ischemic stroke revealing an SLE, in order to analyze the epidemiological, clinical and therapeutic data of these exceptional lesions. A literature review will be presented.

Case report :

A 38-year-old woman, with no notable pathological history, consulted an emergency response for aphasia and rapidly evolving flaccid tetraparesis with impaired consciousness. The patient was intubated and ventilated. Cerebral angio-MRI showed a recent centro-protuberant ischemic lesion with a thrombosis of the basilar trunk and the left vertebral artery (Figure 1).

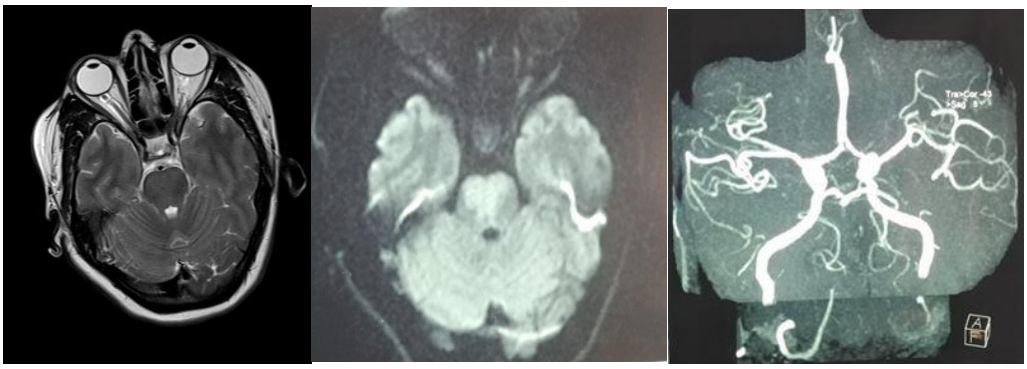

Figure 1 : Cerebral MRI showing the thrombosis of left vertebral artery

The spinal MRI (Magnetic resonance imaging) showed a bilateral lateral medullary hyperintense on T2 wheited images along the cervical and dorsal cord compatible with longitudinal transverse myelitis (Figure 2).

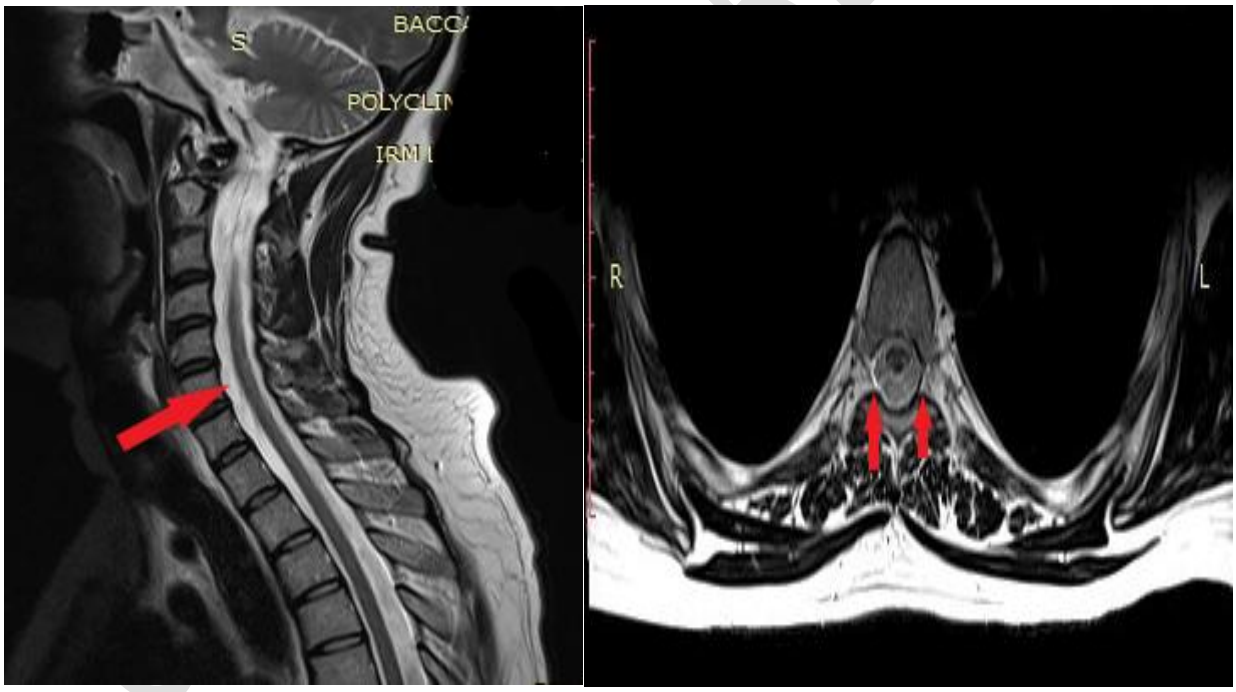

Figure 2 : Spinal angio-MRI showing the cervical and dorsal myelitis.

After a short stay (5 days) in intensive care, the patient's state of consciousness was improved. A locked in syndrome was noted: aphasia, facial diplegia with conservation of the occulomotricity and flaccid tetraplegia
In front of this situation (young patient with ischemic stroke), the search for thrombophilia was negative (anti-phospholipid antibodies, circulating anticoagulant, homocysteinemia, resistance to activated protein C, deficit in protein C and S and anti thrombin III).

An infectious origin of this thrombotic accident has been eliminated in the face of the negativity of the tuberculosis assessment and serologies: Syphilis, HIV, Lyme, HTLV1 and Wright. . A lumbar puncture was indicated but it was refused by the patient.
Optic neuromyelitis was eliminated due to the negativity of anti NMO-IgG antibodies (anti Neuromyelitis Optica type IgG.
We concluded that the SLE is the etiology in front of the presence of 5 criteria of SLICC (SystemicLupus International CollaboratingClinics): Malar rash, anti-nuclear antibodies positive at 1/640, lymphopenia at 900 elements / mm3, pericardial effusion and myelitis.
Despite the rapid introduction of intravenous corticisteroid therapy (1000 mg methylprednisolone (solumedrol ©), 3 days in a row), and cyclophosphamide) infusions (6 monthly boli then 6 quarterly boli) , followed by oral prednisone (1mg / kg / day), as well as motor rehabilitation, our patient retained paraplegia with urinary incontinence.
Curative anticoagulation was not indicated because of the negativity of antiphospholipid antibodies.
Cerebral angio-MRI showed sequelae of bilateral para-sagital pontine ischemic stroke with hemorrhagic stigmata and moderate cerebellar atrophy (Figure 3).
Currently our patient is on prednisone (10 mg / kg / day) and Hydroxychloroquine (6.5 mg / kg / day).

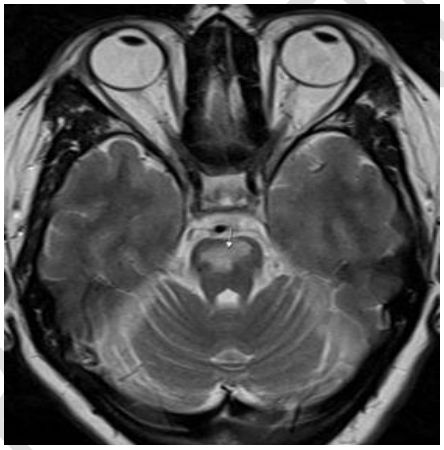

Figure 3 : angio-MRI showing the cerebral sequelae

Discussion :

Central neurological damage during lupus is predominant with serious lesions. Cerebrovascular manifestations constitute one of the most severe attacks that can cause a locked-in syndrome. Transverse myelitis is one of nineteen neuropsychiatric manifestations of SLE, defined by the American College of Rheumatology (ACR) under the term neuropsychiatric lupus (NPSLE). Although the frequency of transverse myelitis during SLE is multiplied by 1000 compared to the general population (5), lupus remains an exceptional etiology of acute transverse myelitis, found in 1.42% of the series of Alvarenga and al (6). A literature review carried out in 2014 reported only 93 cases of lupus myelitis reported over a 50-year period (7).

Lupus myelitis affects both sexes, with no predilection of age. Some pediatric observations have been reported (8).

The circumstances of discovery of lupus myelitis are multiple, ranging from non-specific symptoms such as isolated vomiting (9) to "catastrophic" presentations : tetra or paraplegia with sphincter disorders such as the case of our patient.

The definition of transverse myelitis remains clinical (10), but a radiological definition is currently validated, defining transverse myelitis as a spinal cord lesion covering at least 50% of the transverse surface of the cord. The involvement of at least 3 contiguous vertebral bodies defines longitudinal myelitis, the most serious form of lupus myelopathy. Only about ten cases have been reported in the literature (11).
The inflammatory process in transverse myelitis is often focal and involves the cervical, thoracic or lumbar cord . Whole marrow involvement and disseminated encephalitis have also been described (8,12).
There are two types of lupus myelopathy: gray matter, which is characterized by flaccid paresis of the extremities associated with hyporeflexia and sphincter disorders, and white matter, which is accompanied by spasticity with hyperreflexia and optic neuritis (5,13).
Lupus myelitis can be associated in the context of optic neuromyelitis or Devic disease, a demyelinating neuro-immunological disease, with anti NMO-IgG antibodies (anti NeuromyelitisOptica type IgG), or anti aquaporin-4-IgG (9,14 ,15). The positivity of these antibodies can help differentiate the neurological manifestations of lupus from those of Devic's disease (16).
Treatment is based on the rapid introduction of corticosteroids in high doses and cyclophosphamide as a bolus, offering better neurological recovery (17). This therapeutic approach is validated by the recommendations of the EULAR (European League Against Rheumatism) for the management of the neuropsychiatric manifestations of SLE (18). Early and intensive neurological rehabilitation is an integral part of treatment (19). Systematic anticoagulation for myelitis associated with anti-phospholipid antibodies has not demonstrated any particular beneficial effect, despite the fact that these thrombogenic autoantibodies have been implicated in the pathogenesis of lupus myelitis (20).
In forms associated with Devic's disease, rituximab and cyclophosphamide are effective (16).

Conclusion :

Locked-in syndrome after vascular thrombosis during lupus is associated with a poor vital and functional prognosis. Longitudinal transverse myelitis is a rare manifestation of neurolupus. It can be the inauguration of an SLE. Despite its rarity, the lupus hypothesis must be considered in the face of all myelitis in young women, due to the often reserved functional prognosis.

Treatment is based on the combination of cyclophosphamide and corticosteroids, the rapid initiation of which conditions neurological recovery.

Disclaimer regarding Consent and Ethical Approval:

As per university standard guideline, participant consent and ethical approval have been collected and preserved by the authors

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