Demyelinating Polyneuropathy Complicating Systemic Lupus Erythematosus: A Case Report

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Abstract: Systemic lupus erythematosus (SLE) is an inflammatory disease of unknown cause, characterized biologically by producing multiple autoantibodies, the most characteristics are directed against some kernel components such as deoxyribonucleic acid and native nucleosomes. Neurological manifestations are frequent and polymorphous. They are dominated by central attacks, while peripheral attacks are rarer. We report the case of LS, 34 years old, female, Senegalese, divorced, and followed for years for a SLE, living in Dakar, hospitalized in October 2016 at the Neurological Clinic of FANN National Teaching Hospital, Dakar-Senegal for a demyelinating polyneuropathy complicating Systemic lupus erythematosus (SLE). An electro neuro myogram showed elongation of distal latencies in the lower limbs, lengthening of F waves in the lower limbs, and decreased conduction velocity in the lower limbs. The search for native anti-DNA antibodies was positive. The outcome was fatal with one death on day 41 of his hospitalization.

Keywords: Systemic Lupus Erythematosus, Polyneuropathy, Demyelinating

1. Introduction

Neurological manifestations in systemic lupus erythematosus (SLE) are frequent and polymorphic. They are dominated by central attacks. However, peripheral attacks are rarer, present in 5% of cases. They are represented essentially by polyneuropathies and pure sensorial or sensitivo-motor symmetrical axonal mononeuropathies [1].

A study by S. Elherrar M and al, showed a frequency of 5.59% peripheral neurological involvement in systemic lupus erythematosus with an average age of 37.16 years and a female predominance in 74.2% of cases. The neurological impairment was revealed after an average delay of 37 months of lupus disease. Clinical manifestations of peripheral neurological disease were dominated by sensory disorders in 58% of cases, followed by motor disorders in 29% of cases and amyotrophy which was objectivized in 22.58% of cases and steppage in 19.35% of cases and cranial nerve involvement in 19.35% of cases. In the same study, an association was found between central and peripheral neurological manifestations in the same patient in 41.9% of cases [2].

Demyelinating lupus polyneuropathy is a rare presentation. We report a case of demyelinating polyneuropathy complicating systemic lupus erythematosus in a young adult admitted to the Neurological Clinic of the University Hospital Center of FANN, Dakar-Senegal.

2. Medical Observation

Mrs. LS, 34 years old, divorced, living in Dakar, Senegal was admitted on October 22, 2016 for a motor deficiency of the lower limbs that had settled over 4 days requiring her hospitalization at the Neurological Clinic of the University
Hospital Center of FANN, Dakar-Senegal. She was known as a lupus patient and since 2013 has been attending the dermatology department of the Le Dantec hospital in Dakar, Senegal, and re-hospitalized in 2015 for a second lupus outbreak following a therapeutic rupture. No cases of systemic lupus erythematosus (SLE) have been reported in his family. The clinical examination showed an alteration of the general condition (Stage II) of the WHO with mucosal conjunctiva little colored. There was no fever. A four-paresis predominantly crural with MRC scale at 3/5 on the upper limbs and 0/5 on the lower limbs with hypotonia of the lower limbs, tactile hypoesthesia without metameric level of the lower limbs with saddle anesthesia and Indifferent plantar reflexes. The idiomuscular response was present. The dermatological examination revealed annular disseminated erythema (of the hands and plants of the feet, Fig. 1, 3, 4), psoriasiform and atrophic scarring lesions (at the head) and total alopecia (Fig. 2). In front of this table, the biological tests carried out were positive for Native anti DNA antibodies. The blood count showed: hemoglobin at 7.6 g / dl, hematocrit at 22.2%, mean globular volume at 78 µm³, red blood cells at 2.78.10⁵ / mm³, reticulocyte level at 2, 3%, platelets at 100.10⁵ / mm³, white blood cells at 10.100 10³ / mm³, a sedimentation rate of 50-80 mm / h (1st hour-2nd hour), a CRP at 48 mg/l. Examination of the cerebrospinal fluid showed a protein level at 0.71 g/l and a Glucorachia at 0.34 g/l. Urea was at 0.6 g/l and a 25 mg/l serum creatinine. The 24h proteinuria was not done. Retroviral serology (HIV) was negative. The diagnosis of neurogenic syndrome of the four limbs with a crural predominance was evoked and an electro neuro myogram (ENMG) was performed which showed an extension of the distal latencies of the lower limbs, an elongation of the F waves in the lower limbs and a decreased conduction velocity in the lower limbs. The patient was treated with corticosteroids at a dose of 1mg / kg / day and adjuvants (Calcium, potassium, and gastric protectant), transfusion of whole blood iso group iso rhesus. The motor reeducation was done. The evolution was marked by an aggravation of its state and by the occurrence of an ascites of great abundance. The death will occur on December 3, 2016 or at D41 of its hospitalization

3. Discussion

For some authors, peripheral neurological disease is rare in lupus with a variable frequency depending on the diagnostic criteria used. According to the clinical criteria [3], this frequency is 2.5 to 2.18% of the cases, whereas for the electrophysiological criteria [4] it is of the order of 25 to 30%
of the cases. For Meyer and al, the frequency of lupus polyneuropathy would vary according to ethnic parameters, service recruitment pattern, age and sex [5].

Some others believe that neurological damage is less frequent in the black skin subject than in the Caucasian subject [6]. Peripheral neurogenic involvement during lupus is more an axonal polyneuropathy as shown by a retrospective Moroccan study conducted from 1981 to 2007 [6], which found an average age of patients who was 35 years old with the possibility of other neuropathies such as multiple neuropathy or mono radiculoneuropotic neuropathy and cranial nerve damage in 19.35% of cases, yet in our patient it was a demyelinating polyneuropathy.

For their part, Frigui and al [7], S. Elherrar [2] found an average age of 25.65 years and 37.16 years, respectively, with a female predominance. This data would place our patient in the most affected age group. The absence of cases of lupus demyelinating polyneuropathy reminds us of the small size of the African series which would explain the underestimation of these attacks.

According to Elherrar and al [2], the neurological impairment was revealed after an average duration of lupus disease of 37 months (2 months-26 years), whereas for our patient polyneuropathy was discovered after a disease evolution of 46 months.

S. Ketari and al. [8] had reported in their study the occurrence of sensory disorders such as paresthesia in the sock that preceded by two months the installation of the functional impotence as well as a heaviness of the lower limbs with an electro neuro myogramm which was in favor of a multiple sensitivo motor axonal mononeuropathy that predominated in the lower limbs. In our patient, the sensory disturbances were concomitant with the installation of a functional impotence.

For E. TurkiJaidane and al [1], peripheral neuropathy is often asymptomatic; it often occurs during a relapse and is rather an ischemic than an immunological mechanism. These are usually pure sensorial or sensitivo motor symmetrical axonal polyneuropathies. The presence of anti-phospholipid antibodies (APL) increases the risk of peripheral neuropathies, hence the interest of looking for these antibodies to facilitate the detection of peripheral neurogenic involvement.

The hematological disorders in our case were essentially thrombocytopenia and anemia. This anemia was found in other studies including those of Kambake et al [9], and that of Ka and al [10] who found a frequency of 77%. For Elherrar and al. Hematological or joint damage was associated in 83.87% of cases [2].

In large series, cutaneous signs were present in 68% of the patients with alopecia of the scalp which was found in 43.75% of the cases [9, 10].

The re-hospitalization observed during our patient's illness was also observed by Ka and al [10] in 31.25% of the cases. This would show the difficulty in the medium- and long-term follow-up of a chronic illness in poor countries where the cost of the LSE (medication, balance sheet, purchase) is not negligible. Sometimes financial hardship would be responsible for the non-observance of treatment and loss of sight. This leads to an overall mortality up to 31.25% of cases, as reported in the other african series, especially those of Cameroon by Youmbissi and al [11], South Africa by Jacyk WK and al [12], as well as the study by Mody GM [13].

Studies by Kombate K and al [9], on the other hand, reported generally satisfactory results, namely a favorable outcome in 87.50% of cases in patients with lupus who had neuropathy and who were given early corticosteroids at adequate doses before the installation of kidney failure. Stabilization or even disappearance of manifestations for months or even years was noted.

For Ward MM and al [14], the classic factors of poor prognosis were renal insufficiency, neurolupus, and the male sex. However, other studies have shown that ethnicity is not an important prognostic factor, in contrast sociocultural factors (poverty, precariousness, absence of social security cover) were the main risk factors for the prognosis of systemic lupus [4, 15].

4. Conclusion

Demyelinating polyneuropathie in lupus is rare, and should be systematically investigated in front of any lupus panel in order to provide adequate treatment.

References


