

Severe Axonal Peripheral Polyneuropathy Revealing a Systemic Lupus Erythematosus: About One Case

Mariam Gbané-Koné, Mohamed Diomandé, Boubacar Ouali, Abidou Kawalé Coulibaly, Astrid Nawé Ngandeu, Edmond Eti, Marcel N'zué Kouakou

Department of Rheumatology, University Hospital Center of Cocody, Abidjan, Côte d'Ivoire
Email: Mariamgbane05@yahoo.fr

Received 7 March 2015; accepted 10 April 2015; published 14 April 2015

Copyright © 2015 by authors and Scientific Research Publishing Inc.
This work is licensed under the Creative Commons Attribution International License (CC BY).
<http://creativecommons.org/licenses/by/4.0/>



Open Access

Abstract

Aim: We report a case of acute and severe sensorimotor peripheral polyneuropathy (with a severe motor damage) revealing a lupus. **Case Presentation:** A 48-year-old female patient was interned in rheumatology for a chronic polyarthritis. Four days after her hospitalisation, she was presenting a flask distal and proximal tetraparesia, with rapidly progressive installation. Electromyogram showed severe acute axonal sensorimotor polyneuropathy. The antinuclear antibody was positive as the anti-ds-DNA antibodies. The evolution has been unsatisfactory despite the high-dose corticotherapy and the immunosuppressor. **Conclusion:** Even if it is rare, peripheral neuropathy can be a lupus discovery circumstance.

Keywords

Systemic Lupus Erythematosus, EMG, Neurolupus, Peripheral Polyneuropathy

1. Introduction

Polyneuropathies are defined as a diffuse damage of the peripheral nervous system (PNS), as opposed to unique or multiple neuropathies, characterised by one or many nervous trunks attack(s); or as opposed to polyradiculoneuropathies suggesting an extension of the nervous damages on roots [1].

Systemic lupus erythematosus is a systemic disease, characterised by a multivisceral attack of variable gravity. It occurs preferentially on young woman [2].

The neurological complications are classically considered as the most serious in SLE. They are of variable frequency (40% to 75% of cases) [2]. Central damages are most frequent and are dominated by psychiatric

troubles and convulsive crisis [2] [3].

Peripheral nervous system damage is rarer and noticed in 2% - 30% of cases [2]-[4]. It is rarely revealing the disease [5].

We report a case of severe acute axonal sensorimotor polyneuropathy of the four members revealing a lupus on a 48-year-old female patient. The evolution has been unsatisfactory despite the high-dose corticotherapy and the immunosuppressor.

2. Observation

A 48 years old female patient, interned in the ward for a bilateral polyarthritis, symmetric, touching the bends, hands, wrists, knees, and ankles, with a notion of morning steepness, evolved in a low-grade fever context, with a general body state alteration. This polyarthritis evolved by pushes since one year in a chronic anaemia context. She had a past medical history of pulmonary tuberculosis, a notion of spontaneous abortion.

At the physical exam, the number of painful articulations was 10, there was no joint inflation. There were no cutaneous and mucosal signs (facial erythema, alopecia, oral ulcerations), besides, no visceral damage was noticed. The rest of the exam was without particularity.

Four days after her hospitalisation, the clinical presentation was suddenly deteriorated, the patient was presenting a flask distal and proximal tetraparesia, with rapidly progressive installation; firstly the inferiors members (LSM: 2/5; RSM: 3/5) with paraesthesia's in boot as itch, followed two days after by a superior members damage (IM: 2/5) also with paraesthesia's in gloves. The damage was most marked in distal. It was no respiratory tract damage, no gulp trouble, or cranial nerves damage. Bone and tendinous reflexes were abolished. The cutaneous-plantar reflex was absent. We noticed distal oedemas on the four members. A massive amyotrophy of rapid onset was associated.

Laboratory investigations revealed: an important inflammatory syndrome (ESR 110 mm/1st hr, CRP normal at 8 mg/l). At the hemogram, we had a hypochromic anaemia at 9.7 g/dl. Hepatic balance and glycaemia were normal. Urea at 0.42 g/l; creatininemia at 6.9 mg/l. Muscular enzymes were normal. Viral hepatitis markers were negatives, as the retroviral serology.

The antinuclear antibody were positive (1/1280, speckled aspect) as the anti ds-DNA antibodies (72 UI/ml) and anti-ENA (Anti-Sm = 481 UI/ml, Anti-SSA = 241 UI/ml, Anti-SSB = 16 UI/ml, Anti-URNP = 241 UI/ml).

Anti-CCP were negative as rheumatoid factors. In urine, the 24 hrs urinary protein was positive (2.60 g/24H).

Electromyogram showed severe acute axonal sensorimotor polyneuropathy of the four members: with a motor amplitude collapse, the preservation of the speed of conduction and an absence of conduction's blocs. The Doppler echography, and the pulmonary radiography were normal. The hands, wrists, and ankles radiographies did not objective erosions or joints destructions. The lupus diagnosis with neurologic complications (axonal PPN of members) and kidneys complications has been established. The therapy was constituted by bolus of (240 mg) methylprednisolone, at the first days, associated by cyclophosphamide cures (Endoxan). The corticoid switch per os was 1 mg/kg/day. Endoxan was introduced at 600 mg/m² in bolus, one time monthly. A functional reeducation has been concomitantly prescribed.

The evolution was not spectacular despite this treatment, it is important to notice that Endoxan cures just start one month after the diagnosis, and also have not been regular because of financial difficulties of the patient (no health insurance).

The 9th month of treatment, functionally, the evolution was unsatisfactory, she received just 4 Endoxan cures, the corticotherapy was maintained until this day at 1mg/kg, there was no diabetes inducted by corticosteroids. The reeducation has not been well followed, we can continue to note the deficit of the 4 members and the amyotrophy (the patient can stand up with help, the walking is not possible, the left hand is falling).

3. Discussion

The particularity of this observation is the inaugural character of PPN revealing the SLE (besides the joint damages) and the motor damage severity. Indeed, in most of cases, polyneuropathy arises during a lupus evolution, in general rule, many years after the diagnosis [3]. It can sometimes arise during a clinical or biological lupic push. Rarely, it can be isolated and revealing [5] [6]. Neuropathy was revealing of lupus in our patient.

Rafai [3] reported a case of a 48 years old woman presenting a sensorimotor polyneuropathy with severe motor involvement complicating lupus associated with antiphospholipids antibodies. Outcome was good after cyc-

lophosphamid pulse.

PPN can be associated to a central damage [3], it can also product itself with an absence of central neurological manifestations [6]-[8]; that was the case in our female patient, she did not have CNS signs.

The peripheral damages of lupus are generally axonal symmetric polyneuropathies; multiple mononeuropathies, or cranial nerves damages; exceptionally demyelinating polyneuropathy or polyradiculoneuritis like Guillain-Barre [5] [9]-[12].

A lot of mechanisms have been cited to explain the peripheral damage during lupus. The most important is an ischemic vascular mechanism, by vas a nervorum vascularitis or by micro thrombi linked to antiphospholipids antibodies [2] [4] [6].

The other proper mechanisms are: immunologic cause by a direct aggression by antibodies, entraining a destruction of the peripheral nerve component [6]; or at the end a metabolic disorder [5] [6] [8]. On our patient, EMG made it possible to the diagnosis of severe axonal peripheral neuropathy. Electrophysiology is essential in the diagnosis, it permit to confirm a damage of the PNS, precise the type and the physiopathological mechanism [1]. The electroneuromyographic exam (ENMG) will permit to order the neuropathy in function of the causal mechanism: axonopathies, myelinopathies or neuronopathies [1]. For our patient, the damage was acute sensorimotor. Cases of pure sensitive neuropathy have been described in lupus [13]-[15].

In absence of controlled studies, the neurolupus therapeutic aspects are widely based on team experiences [2]-[4].

High doses corticosteroids are part of the treatment, associated or not to immunoglobulins, immunosuppressors, to plasmatic exchanges, mostly in severe forms [2]-[4].

The evolution of these neuropathies is unpredictable, going to a response to the treatment [5] at a bad therapeutic response [8]. Relapses are possible. In our case, the evolution has not been spectacular despite the association of corticoids and immunosuppressors.

4. Conclusion

Even if it is rare, peripheral neuropathy can be a lupus discovery circumstance. We must have it in mind, in front of all members' deficit of young woman associated to polyarthralgias and to an important inflammatory syndrome.

References

- [1] Stojkovic, T. (2006) Les neuropathies périphériques: Orientations et moyens diagnostiques. *La Revue de médecine interne*, **27**, 302-312. <http://dx.doi.org/10.1016/j.revmed.2005.10.018>
- [2] Ahbeddou, N., Benomar, A., Rasmouni, K., Quessar, A., Ouhabi, H., Ait Ben Haddou, E. and Yahyaoui, M. (2010) Thrombose veineuse cérébrale et polyradiculonévrite aigue révélant un lupus érythémateux disséminé. *Revue Neurologique*, **166**, 458-463. <http://dx.doi.org/10.1016/j.neurol.2009.09.005>
- [3] Rafai, M.A., Fadel, H., Boulaajaj, F.Z., Gam, I., El Moutawakkil, B., Karkouri, K., Hakim, M. and Slassi, I. (2007) Neuropathie périphérique au cours du lupus érythémateux disséminé avec vascularite épineurale et anticorps phospholipides. *Revue Neurologique*, **163**, 103-106. [http://dx.doi.org/10.1016/S0035-3787\(07\)90363-X](http://dx.doi.org/10.1016/S0035-3787(07)90363-X)
- [4] Ha-ou-noua, F.Z., Dehbia, S., Zahlanea, M., Kissanib, N. and Essaadouni, L. (2014) Polyradiculonévrite aiguë révélant un lupus érythémateux systémique: Une présentation inhabituelle d'évolution fatale. *La Revue de Médecine Interne*, **35**, 65-67. <http://dx.doi.org/10.1016/j.revmed.2012.11.010>
- [5] Karmochkine, M., Bironne, P., Gherardi, R., Jarrousse, B., Glowinski, J. and Guillevin, L. (1993) Neuropathie périphérique avec vascularite épineurale et myélite transverse au cours du lupus érythémateux disséminé. *Annales de Médecine Interne*, **144**, 145-147.
- [6] Sridhar, A.V., Gosalakkal, J., Pye, I.F. and Houtman, J. (2004) Peripheral Neuropathy; A Rare Complication of Paediatric Systemic Lupus Erythematosus. *European Journal of Paediatric Neurology*, **8**, 253-256. <http://dx.doi.org/10.1016/j.ejpn.2004.04.004>
- [7] Campello, I., Almarcegui, C., Velilla, J., Hortells, J.L. and Oliveros, A. (2001) Peripheral Neuropathy in Systemic Lupus Erythematosus. *Revue Neurologique*, **33**, 27-30.
- [8] Harel, L., Mukamel, M., Brik, R., Blau, H. and Straussberg, R. (2002) Peripheral Neuropathy in Paediatric Systemic Lupus Erythematosus. *Revue Neurologique*, **27**, 53-56. [http://dx.doi.org/10.1016/S0887-8994\(02\)00377-6](http://dx.doi.org/10.1016/S0887-8994(02)00377-6)
- [9] Ainiala, H., Loukkola, J., Peltola, J., Korpela, M. and Hietaharju, A. (2001) The Prevalence of Neuropsychiatric Syn-

- dromes in Systemic Lupus Erythematosus. *Neurology*, **57**, 496-500. <http://dx.doi.org/10.1212/WNL.57.3.496>
- [10] Brey, R.L., Holliday, S.L., Saklad, A.R., *et al.* (2002) Neuropsychiatric Syndromes in Lupus: Prevalence Using Standardized Definitions. *Neurology*, **58**, 1214-1220. <http://dx.doi.org/10.1212/WNL.58.8.1214>
- [11] Aït Benhaddou, E., Birouk, N., El Alaoui-Faris, M., *et al.* (2003) Polyradiculonévrite aiguë de type Guillain-Barré révélant un lupus érythémateux aigu disséminé. Étude de deux cas et revue de la littérature. *Revue Neurologique*, **159**, 300-306.
- [12] Bruns, A. and Meyer, O. (2006) Manifestations neuropsychiatriques du lupus érythémateux disséminé. *Revue du Rhumatisme*, **73**, 1293-1300. <http://dx.doi.org/10.1016/j.rhum.2006.05.016>
- [13] Maisonobe, T., Denys, V. and Amoura, Z. (2009) Neuropathies sensitives et maladies auto-immunes: Stratégie diagnostique à propos d'une observation. *Revue Neurologique*, **165**, S70-S76. [http://dx.doi.org/10.1016/S0035-3787\(09\)73951-7](http://dx.doi.org/10.1016/S0035-3787(09)73951-7)
- [14] Omdal, R., Loseth, S., Torbergsen, T., *et al.* (2001) Peripheral Neuropathy in Systemic Lupus Erythematosus—A Longitudinal Study. *Acta Neurologica Scandinavica*, **103**, 386-391. <http://dx.doi.org/10.1034/j.1600-0404.2001.103006386.x>
- [15] Goransson, L.G., Tjensvoll, A.B., Herigstad, A., Mellgrn, S.I. and Omdal, R. (2006) Small-Diameter Nerve Fiber Neuropathy in Systemic Lupus Erythematosus. *Archives of Neurology*, **63**, 401-404. <http://dx.doi.org/10.1001/archneur.63.3.401>