Clinical Neurology and Neuroscience
2021; 5(4): 78-81
http://www.sciencepublishinggroup.com/j/cnn
doi: 10.11648/j.cnn.20210504.12
ISSN: 2578-8922 (Print); ISSN: 2578-8930 (Online)

Axonal Polyneuropathy as Initial Presentation of Lupus in a 15-year-old Male Teenager "Case Report"

Paul-Cedric Mbonda[1, *], Grâce Nkoro[1], Daniel Massi Gams[2], Yannick Fogang[3], Callixte Kuate[1]

[1]Faculty of Medicine and Biomedical Sciences, University of Yaounde I, Yaounde, Cameroon

[2]Faculty of Medicine and Pharmaceutical Sciences, University of Buea, Buea, Cameroon

[3]Faculty of Medicine and Pharmaceutical Sciences, University of Dschang, Dschang, Cameroon

Email address:

mbondapaul@gmail.com (Paul-Cedric M.)

[*]Corresponding author

To cite this article:

Paul-Cedric Mbonda, Grâce Nkoro, Daniel Massi Gams, Yannick Fogang, Callixte Kuate. Axonal Polyneuropathy as Initial Presentation of Lupus in a 15-year-old Male Teenager "Case Report". *Clinical Neurology and Neuroscience.* Vol. 5, No. 4, 2021, pp. 78-81.
doi: 10.11648/j.cnn.20210504.12

Received: July 17, 2021; **Accepted**: August 16, 2021; **Published**: October 19, 2021

Abstract: Systemic lupus erythematosus (SLE) is a chronic autoimmune systemic disease, of the connectivitis group. This condition particularly affects young women between the ages of 20 and 40. Several systems are affected during the disease including the nervous system where central damage is more described than peripheral damage. We report here the case of a 15 year old male teenager with systemic lupus erythematosus whose initial clinical manifestation was acute inflammatory axonal polyneuropathy. This diagnosis was made on the basis of allodynia predominantly in gloves and socks with clinical examination of bilateral peripheral neurogenic syndrome predominant in both lower limbs all associated with fever. The electroneuromyogram showed signs of sensory and motor impairment (more marked in sensory) of the axonal type in both lower limbs, compatible with polyneuropathy. The patient developed several skin lesions such as erythematous and scaly patches on the extremities, and purpuric macules of the palms of hands. All the antibodies tested in the blood came back positive and the skin biopsy described a proliferation of vascular capillaries with a fibrous and myxoid wall, dissociated by inflammatory cells, suggesting inflammatory involvement. Under treatment with hydroxychloroquine and corticosteroids, the patient presented a marked improvement in the general condition as well as on the functional level with regression of sensory and motor disorders.

Keywords: Polyneuropathy, Lupus, Teenager, Case Report

1. Introduction

Systemic lupus erythematosus (SLE) is a chronic autoimmune systemic disease, of the connectivitis group, which has several manifestations which varies among individuals. This condition particularly affects young women between the ages of 20 and 40 [1]. Several systems are affected during the disease (cardiovascular, renal, hematological, respiratory, locomotor, integumentary, and immune) including the nervous system where central damage is more described than peripheral damage [2, 3] In fact, systemic inflammation can lead to aberrant brain-resident immune cell activation, leakage of the blood^{-} brain barrier, and the production of circulating antibodies that cross-react with brain antigens, several manifestations such as: neuropsychiatric disorders, headache, aseptic meningitis, convulsions, abnormal movements, stroke, confusions have been described [4, 5]. In the literature, damage to the peripheral nervous system is less described, these usually occur after a diagnosis of systemic lupus erythematosus has been made and very few are found in young subjects [6, 7]. We report here the case of a patient with systemic lupus erythematosus whose initial clinical manifestation was acute inflammatory axonal polyneuropathy.

2. Main Body

2.1. Patient Information

We report the clinical case of a 15-year-old teenager, who came to consult for dysesthesia of the lower limbs which has

progressed for 1 month, with functional impotence of both lower limbs, all associated with a fever. He had no known chronic condition, no other known medical or surgical history.

2.2. Clinical Fidings

Clinical examination found superficial sensitivity disturbances such as allodynia, particularly in gloves and socks, neuropathic pain (7/10 at DN4) more intensive in the extremities of the lower limbs. In the lower limbs, muscle strength was rated at 4/5 on the proximal muscles and 2/5 on the distal muscles bilaterally. The achilles and patellar reflexes were abolished. Sensitivity examination revealed tactile anesthesia (coarse and fine tact) as well as the abolition of kinesthesia (sense of position of limb segments) on both lower limbs. In the upper limbs, muscle strength was normal proximally and rated at 4/5 for distal muscles symmetrically, ulno-pronator and radial reflexes were normal, and symmetrical. Tactile sensitivity was reduced (coarse tact) on the extremities, and deep sensitivity (kinesthesia, pallesthesia) retained. Trophic disorders such as dryness of the skin more marked on the elbows and ankles were found, but there was no muscular atrophy or edema. The examination of the cardiovascular and respiratory systems was unremarkable.

2.3. Diagnostic Assessment

With these signs of peripheral neurogenic damage to the lower and upper limbs, we performed an electroneuromyogram (ENMG) which showed bilateral axonal-type damage to the lower limbs with decrease in sensory potentials: amplitudes of 4.3µV (normal > 10µV) and 1.6µV (normal > 5µV) respectively on the sural and musculocutaneous right nerves. A decrease in motor potentials was also noted with an amplitude of 1.8 mV for external popliteal sciatic (normal > 3 mV) but the distal motor latencies preserved. The internal popliteal sciatic was not stimulable. Ultimately, we could note a collapse of conduction velocities (almost zero on the sensitive and motor trunks of the lower limbs), symmetrical, predominantly on sensitivity and only on the lower limbs, distal motor latencies, amplitudes and conduction velocities were preserved in the upper limbs (See figures 1 and 2).

The cerebrospinal fluid examination returned to normal (clear appearance, cytology with two cells per mm3, protein 0.40g/l, glucose 0.62g/l, chloride 121.60 mEq/l). The serologies for HIV, hepatitis B and C came back negative. C Reactiv Protein was positive and very high at 122 mg/l. The complete blood count showed anemia at 10.4 g/dl hemoglobin, a hematocrit level of 30.2, a mean corpuscular volume of 97fl; a mean corpuscular hemoglobin concentration of 34 with 9,700 leukocytes and 319,000 platelets. We found no kidney damage (urea 0.39 g/l. Creatinine 9.6 mg/l).

One week after the onset of symptoms, the patient developed several skin lesions such as erythematous and scaly patches on the extremities, and purpuric macules of the palms of hands (See figures 3 and 4).

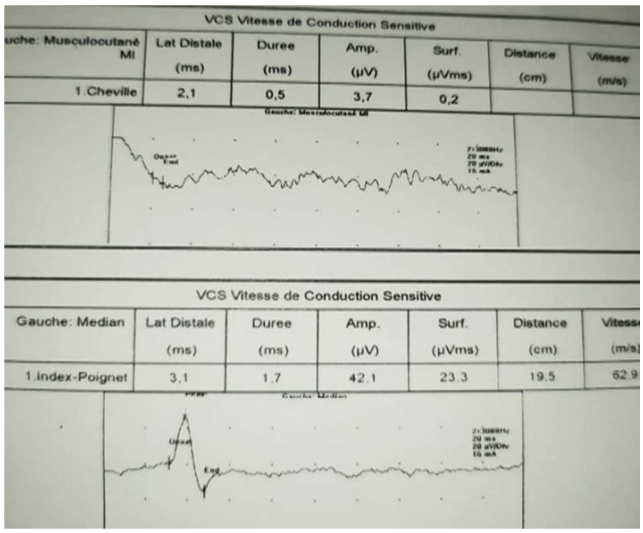

Figure 1. *Decrease in sensory amplitude of the musculocutaneous nerve with collapse of sensory conduction velocity. Sensory amplitude and speed retained on the median nerve.*

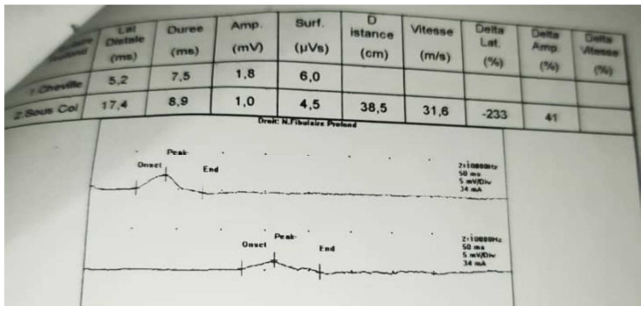

Figure 2. *Decreased motor amplitude in external popliteal sciatic nerve, with collapse of motor conduction velocity.*

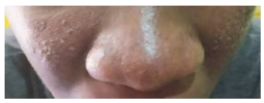

Figure 3. *Erythematous and scaly plaques of the face.*

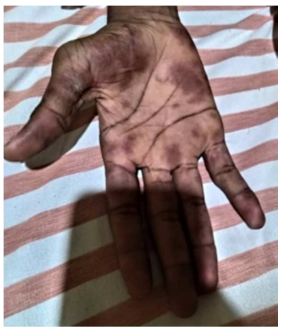

Figure 4. *Purpuric macules of the palms of hands020-12-22-12-03-47.*

The skin biopsy described a proliferation of vascular capillaries with a fibrous and myxoid wall, dissociated by inflammatory cells, suggesting inflammatory involvement.

Faced with these skin lesions, we then suspected a systemic disease and requested native anti-DNA antibodies, antinuclear antibodies (>28), anti SSA antibodies (>241), anti SSB (>417), SLE-specific anti-Sm (>481), anti -RNP (>241),

that all were positive.

Faced with all these elements, we made the diagnosis of systemic lupus erythematosus.

2.4. Therapeutic Intervention

As management, the patient received prednisone at 1 mg/kg/day for 2 months with reduction to 0.5 mg/kg/day currently, and hydroxychloroquine at 400 mg/day since 4 months (treatment in progress).

2.5. Follow-up and Outcomes

We observed a clear improvement in the general condition, in sensitivity with a regression of neuropathic pain (DN4 score from 7/10 to 3/10), a regression of allodynia, evolution from tactile anesthesia in socks to tactile hypoesthesia, kinesthesia of the toes has normalized, all this after 2 weeks of treatment. After 1 month of treatment we observed an improvement on the motor level (the muscle strength in the distal region went from 4/5 to 5/5 in the upper limbs and 2/5 to 4/5 in the lower limbs), the Achilles reflexes initially abolished evolved to a rating of 2+. The patient is also currently doing motor physiotherapy sessions.

3. Discussion

Peripheral nervous system damage (polyneuropathy, mononeuropathy, myasthenia gravis, cranial nerve palsy, acute inflammatory demyelinating polyradiculoneuropathy) can be found in lupus, and in the literature they are found in 1.5% to 15% of cases [8, 9]. Polyneuropathies are the most frequent damage to the peripheral nervous system, according to several authors [10], and are more common in women [11]. These damages of the peripheral nervous system very often occur several years after the diagnosis of lupus has been made [4] and most often affects people over 30 years of age [8, 11], they are frequently found when the patient also has damage to the central nervous system [10]. What makes the particularity of this case, where our patient was a young boy of 15 years whose initial manifestation of lupus was a polyneuropathy. This diagnosis was made on the basis of allodynia predominantly in gloves and socks with clinical examination of bilateral peripheral neurogenic syndrome predominant in both lower limbs. The electroneuromyogram showed signs of sensory and motor impairment (more marked in sensory) of the axonal type in both lower limbs, compatible with polyneuropathy according to the criteria of the American Academy of Neurology [12].

The diagnosis of SLE was made on the basis of clinical criteria: fever, skin lesions (maculopapular rash), non-scarring alopecia, peripheral neuropathy in the absence of other causes, and biological: hemolytic anemia, antinuclear antibody titre higher than the laboratory standard, Anti-native DNA antibodies higher than the laboratory standard, presence of an antibody to the Sm antigen. The definite diagnosis of SLE has been retained according to the American College of Rheumatology [13].

Wang et al in a study conducted on 4924 patients with systemic lupus erythematosus, found polyneuropathies particularly in patients with an advanced form of lupus, and in 0.1% of cases the polyneuropathy appeared before lupus [9]. Fever is one of the clinical signs most frequently associated with polyneuropathies in lupus, and the biology very often finds the positivity of anti-Sm antibodies as described in our patient [8]. The analysis of cerebrospinal fluid returns abnormal most often in acute inflammatory demyelinating lesions (Guillain-Barré like syndrome) according to several authors [14, 15]. Electroneuromyographic examination most often finds a sensory-motor axonal polyneuropathy, with a predominance of sensitivity, and impaired conduction velocities most often linked to axonal damage [8] as found in our patient.

Several molecules are used in the management of lupus: hydroxychloroquine, corticosteroids (cortisone, prednisone, methylprednisone), high doses of aspirin, non-steroidal anti-inflammatory drugs, immunosuppressants and immunomodulators, particularly in refractory cases. Several studies are still underway concerning biological therapies for lupus [14].

Our patient's progress was very satisfactory under treatment with hydroxychloroquine combined with prednisone (which are first-line molecules and more accessible in our context), the literature describes good functional recovery under treatment [15, 16].

4. Conclusion

Systemic lupus erythematosus is an inflammatory disease most commonly affecting the female sex and having well-defined diagnostic criteria. Peripheral nervous system damage in systemic lupus erythematosus are not very frequent, and little described as initial manifestation. This clinical case of a young teenager male who had a polyneuropathy as an initial manifestation shows the multiple forms of inflammatory diseases and should prompt us to carry out an inflammatory assessment in front of any young subject with a clinical picture of polyneuropathy.

5. Recommendations

At the end of this work, we recommend in any case of damage of the peripheral nervous system in the young subject, to make a clinical examination of all the systems, and to carry out a complete inflammatory assessment, in order to make an accurate diagnosis and start the management the earliest.

Competing Interests

All the authors do not have any possible conflicts of interest.

References

[1] Rahman A, Isenberg DA, Systemic lupus erythematosus, N Eng J Med. 2008; 358: 929-939.

[2] Alessandra Bortoluzzi, Ettore Silvagni, Federica Furini, Matteo Piga, Marcello Govoni. Peripheral nervous system involvement in systemic lupus erythematosus: a review of the evidence. Clin Exp Rheumatol. 2019; 37 (1): 146-155.

[3] Jonathan Y Streifler, Yair Molad. Connective tissue disorders: systemic lupus erythematosus, Sjögren's syndrome, and scleroderma. Handb Clin Neurol. 2014; 119: 463-73.

[4] Bendorius M, Po C, Muller S, Jeltsch-David H. From Systemic Inflammation to Neuroinflammation: The Case of Neurolupus. Int J Mol Sci. 2018 Nov 13; 19 (11): 3588.

[5] McGlasson S, Wiseman S, Wardlaw J, Dhaun N, Hunt DPJ. Neurological Disease in Lupus: Toward a Personalized Medicine Approach. Front Immunol. 2018 Jun 6; 9: 1146.

[6] Cavallasca JA, Cousillas JE. Numbness and Weakness in a Patient with Systemic Lupus Erythematosus. Am J Med. 2021 Jan; 134 (1): e53-e54.

[7] Xiangling Li, Yanqiang Wang. Systemic Lupus Erythematosus with Acute Inflammatory Demyelinating Polyneuropathy: A Case Report and Review of the Literature. Case Reports. J Clin Med Res. 2016 Jul; 8 (7): 555-9.

[8] R Jasmin, S Sockalingam, L P Ramanaidu, K J Goh. Clinical and electrophysiological characteristics of symmetric polyneuropathy in a cohort of systemic lupus erythematosus patients. Lupus. 2015; 24 (3): 248-55.

[9] Wang Xianbin, Wang Mingyu, Xu Dong, Li Huiying, Xu Yan, Zhang Fengchun, Zeng Xiaofeng. Peripheral neuropathies due to systemic lupus erythematosus in China. Medicine (Baltimore). 2015; 94 (11): e625.

[10] Brandusa Florica, Ellie Aghdassi, Jiandong Su, Dafna D Gladman, Murray B Urowitz, Paul R Fortin. Peripheral neuropathy in patients with systemic lupus erythematosus. Semin Arthritis Rheum. 2011; 41 (2): 203-11.

[11] Pilar Toledano, Ramón Orueta, Ignasi Rodríguez-Pintó, Josep Valls-Solé, Ricard Cervera, Gerard Espinosa. Peripheral nervous system involvement in systemic lupus erythematosus: Prevalence, clinical and immunological characteristics, treatment and outcome of a large cohort from a single centre. Autoimmun Rev 2017; 16 (7): 750-55.

[12] J. D. England, G. S. Gronseth, G. Franklin, G. T. Carter, L. J. Kinsella, J. A. Cohen, A. K. Asbury, K. Szigeti, J. R. Lupski, N. Latov, R. A. Lewis, P. A. Low, M. A. Fisher, D. N. Herrmann, J. F. Howard, G. Lauria, R. G. Miller, M. Polydefkis, A. J. Sumner. Practice Parameter: Evaluation of distal symmetric polyneuropathy: Role of laboratory and genetic testing (an evidence-based review). Neurology. 2009; 1 (1): 5-13.

[13] Martin Aringer, Nicolai Leuchten, Sindhu R Johnson. New Criteria for Lupus. Curr Rheumatol Rep. 2020; 22 (6): 18.

[14] J K Presto, E Z Hejazi, V P Werth. Biological therapies in the treatment of cutaneous lupus erythematosus. Lupus. 2017; 26 (2): 115-118.

[15] Coomes EA, Haghbayan H, Spring J, Mehta S. Fulminant Guillain-Barre syndrome in a patient with systemic lupus erythematosus. BMJ Case Rep. 2019 Jan 14; 12 (1): bcr-2018-226634.

[16] Sodavarapu S, Rudrangi R, Hoffman J. Rare case of bilateral wrist and foot drop from SLE-related vasculitic polyneuropathy. BMJ Case Rep. 2020 Jan 15; 13 (1): e232890.