AXONAL POLYNEUROPATHY AS INITIAL PRESENTATION OF LUPUS IN A 15-YEAR-OLD MALE TEENAGER “CASE REPORT”

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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 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 based on alldynia 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. In addition, the patient developed several antibodies tested in the blood that returned positive. The skin biopsy described the 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

Introduction

Systemic lupus erythematosus (SLE) is a chronic autoimmune systemic disease of the connectivitis group, which has several manifestations which vary 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, haematological, respiratory, locomotor, integumentary, and immune), including the nervous system, where central damage is more described than peripheral damage [2, 3].

We report the case of a patient with systemic lupus erythematosus whose initial clinical manifestation was acute inflammatory axonal polyneuropathy.

Patient and Observation

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 and no other known medical or surgical history.

Clinical findings

Clinical examination found superficial sensitivity disturbances such as alldynia, particularly in gloves and socks, and neurogenic 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 anaesthesia (coarse and fine tact) as well as the abolition of kinesthesia (sense of the 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) was 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 oedema. The examination of
the cardiovascular and respiratory systems was unremarkable.

**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 a 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 were 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 was 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 Reactive Protein
was positive and very high at 122 mg / l. The complete blood
count showed anaemia at 10.4 g/dl haemoglobin, a hematocrit
level of 30.2, a mean corpuscular volume of 97fl, and a mean
corpuscular haemoglobin 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 the sensory amplitude of the musculocu-
taneous nerve with the collapse of sensory conduction velocity.
Sensory amplitude and speed retained on the median nerve.

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

Figure 3 Erythematous and scaly plaques of the face.

Figure 4 Purpuric macules of the palms of hands.
The skin biopsy described the 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. We observed 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 diagnosed systemic lupus erythematosus.

**Therapeutic Intervention**

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

**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 anaesthesia 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.

**Discussion**

Peripheral nervous system damage (polyneuropathy, mononeuropathy, myasthenia gravis, cranial nerve palsies, acute inflammatory demyelinating polyradiculoneuropathy) can be found in lupus. In the literature, they are found in 1.5% to 15% of cases [4, 5]. Polyneuropathies are the most frequent damage to the peripheral nervous system, according to several authors [6], and are more common in women [7]. These damages to 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 [4, 7]; they are frequently found when the patient also has damage to the central nervous system [6]. So what makes the particularity of this case, where our patient was a young boy of 15 years whose initial manifestation of lupus was polyneuropathy? This diagnosis was made based on 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 American Academy of Neurology [8].

The diagnosis of SLE was made based on clinical criteria: fever, skin lesions (maculopapular rash), a non-scarring alopecia, peripheral neuropathy in the absence of other causes, and biological: hemolytic anaemia, 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. However, according to the American College of Rheumatology [9], the definite diagnosis of SLE has been retained.

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. In 0.1% of cases, polyneuropathy appeared before lupus [5]. Fever is one of the clinical signs most frequently associated with polyneuropathies in lupus. Biology often finds the positivity of anti-Sm antibodies, as described in our patient [4]. The analysis of cerebrospinal fluid returns abnormal most often in acute inflammatory demyelinating lesions (Guillain- Barré like syndrome), according to several authors [10]. 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 [4] 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. In addition, several studies are still underway concerning biological therapies for lupus [10].

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 [7, 10].

**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 is not frequent, and little is described as an initial manifestation. This clinical case of a young teenage male with polyneuropathy as an initial manifestation shows the multiple forms of inflammatory diseases and should prompt us to perform an inflammatory assessment in front of any young subject with a clinical picture of polyneuropathy.

**Competing Interests**

Authors have no financial, political, personal, religious, ideological, academic, intellectual, commercial or any other conflicts of interest to declare in relation to this manuscript.

**Authors’ Contribution**

PCM and GN examined the patient in the hospital; PCM did all his follow-ups and wrote the first draft of this case report. PCM, GN, DGM, YFF and CTK revised subsequent versions and approved the final article.

**Patient perspective**

According to the patient and his family, the treatment (still in progress) is really effective in view of its improvement but has side effects such as weight gain.

**Informed consent**

The authors had the informed consent of the patient’s parents.

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