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Advance directive in a young patient: Palliative care in the context of grade 3 lupus nephritis

Diretiva antecipada de vontade em paciente jovem: Cuidados paliativos no contexto de nefrite lúpica grau 3

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ABSTRACT

Systemic lupus erythematosus is a systemic autoimmune disease that compromises several organ systems, being capable of reducing the quality of life of patients in many cases. Early diagnosis is associated with poor prognosis since the period of disease activity is longer. Lupus nephritis is one of the consequences that can arise, and the chronic kidney disease that follows becomes terminal in 10% of cases. The present study reports the case of a young lupus patient with grade 3 lupus nephritis who, due to the scarcity of resources that could alleviate her suffering and restore her quality of life, chose not to receive advanced life support in future hospitalizations.

Keywords: Systemic lupus erythematosus, Advance directive, Lupus nephritis.

INTRODUCTION

Systemic lupus erythematosus (SLE) is characterized as an autoimmune disease with systemic repercussions, with damage concentrated in organs such as the skin, lungs, heart, joints, central nervous system, and kidneys. Both sexes, all races - with lower prevalence in the white race - and age groups, including children, are subject to the manifestations of this disease if it develops. Without proper treatment, complications arise as a result of the deterioration of the functions of the target organs, which can evolve with the death of the patient [1].

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The incidence of systemic lupus erythematosus in children is significantly lower than in adults, with prevalences in the order of 0.3 to 0.9 per 100,000 children, compared with 3.3 to 8.8 per 100,000 adults. In childhood, SLE usually manifests itself in the transition to early adolescence, around 10 to 12 years of age – in this age group there is a concentration of about 10 to 20% of diagnoses. In cases with early onset of the disease, there is a strong correlation with worse prognosis, since the organism is subject to organic changes of SLE for a longer period [2].

Among the complications that can arise in organs and systems due to the manifestations of the disease, renal complications are the most commonly found in patients. About 50 to 75% of patients diagnosed with SLE will develop chronic kidney disease (CKD) by the end of life, and 10% of these will develop the disease in its dialysis/terminal form [2]. Lupus nephritis is usually diagnosed within the first 5 years after the diagnosis of SLE, and is therefore one of the first manifestations of target organ involvement to develop [3].

Other manifestations that may occur due to SLE, less frequently, but with very important repercussions, are neuropsychiatric manifestations. There may be repercussions on the central nervous system, autonomic, psychiatric disorders, diffuse and focal disorders. Much less frequent than lupus nephritis, lupus psychosis affects about 2.5% of patients with SLE. Symptoms include delusions of grandeur, paranoids, visual and auditory hallucinations, but there are many gaps to be answered about the single psychiatric question, since they are manifestations that are difficult to report due to their low incidence [4].

There is no consensus on how SLE mechanisms can, in fact, alter the nervous system to the point of causing hallucinations and delusions, and recent research shows that about 30% of cases of neuropsychiatric events in patients with SLE are a direct result of disease activity, which calls into question the occurrences in the rest of the patients [5].

The objective of this scientific text is to report a clinical case of a young patient, 18 years old, diagnosed with SLE 10 years ago, in outpatient treatment in a tertiary hospital in the interior of the state of São Paulo, who, due to the unfavorable evolution of the disease, already with severe renal involvement, chose to refuse future major interventions. maintaining only symptomatic treatment for manifestations of the disease. The patient's wishes were heard by the clinical staff, discussed among the professionals and complied with via advance directive.

METHODOLOGY

Review of electronic medical records and literature review in databases such as PubMed, Lilacs and Scielo. The present study was approved by the internal ethics committee of the hospital where the care was provided, following the guidelines of the National Research Ethics Council (CONEP).

CLINICAL CASE REPORT

An 18-year-old female patient diagnosed with systemic lupus erythematosus more than 10 years ago was admitted to the emergency room, referred to a city in the region, after dyspnea, prostration, asthenia, and fatigue. Laboratory tests showed severe leukopenia, thrombocytopenia and anemia. The patient was hospitalized and transfusion with packed red blood cells was initiated. The patient denied any previous bleeding, and there were no hemorrhages during the hospitalization period.

As this is a long-term patient, in outpatient treatment for more than 10 years, undergoing drug therapy for lupus and depression, in view of the laboratory pattern presented, the diagnostic hypothesis of bone marrow aplasia secondary to the use of medication was inquired about, and a rheumatology opinion was requested to follow up the case. Analyzing the case, it was decided to perform pulse therapy with methylpredinisolone for 3 days, to be started on the following day, and to maintain the oral medication after this period.

The next morning, when the first pulse with methylpredinisolone was about to be started, the patient showed resistance to the treatment, to which she had already been submitted a little more than 4 months ago. She expressed a desire to escape from the hospital, which drew the attention of the intensive care unit (ICU) team. Guidance was given to the patient, who calmed down and did not receive the medication. A psychiatry opinion was requested, due to suspicion of lupus psychosis, in addition to a discussion in conjunction with other specialties to enable a possible advance directive of will to meet the patient's determination in case of being removed from psychoticism.

The patient in question had a very early diagnosis of SLE, and because of this she had already been in treatment for more than a decade, even though she was so young. As a result of baseline lupus, grade 3 lupus nephritis developed, which triggered marked loss of renal function and arterial hypertension. Parallel to these diagnoses, there was the discovery of an aortic valve disease and mild mitral regurgitation. Due to the exhausting routine of outpatient treatment, the displacements from her city of origin and the fact that she was away from her mother during these periods, the patient developed a depressive condition, which culminated in an attempt at self-extermination.

At the time, she was admitted to the psychiatric ward of the hospital, and when she was discharged, the patient began to use antidepressants. There was always good adherence to the proposed treatments, however, in eventual consultations where the mother could attend, she reported drug addiction and alcoholism on the part of her daughter.

Questioned about her decision to be discharged without pulse therapy, and not to receive advanced life support in subsequent hospitalizations, the patient maintained her position. The consultation with psychiatry ruled out lupus psychosis, and an appointment with the hospital's palliative care team was requested. The patient was aware of the severity of her clinical condition and the impact of her decision on the course of treatment and also on her prognosis of life. A meeting was scheduled between specialties and family members to prepare an advance directive of will (DAV).

All doubts of both the family and the patient were clarified. All attending physicians issued their opinions and, regardless of their personal perceptions or even therapeutic expectations as professionals, respected the patient's position regarding her prognosis. As a result, 4 days after her hospitalization, the patient had her advance directive formalized, where she was agreed about hospital discharge, maintenance of SLE therapy, antidepressant therapy, and refusal in situations where advanced life support was required in cases of future hospitalizations.

RESULTS

After researching the patient's medical records, we evidenced the complexity of her case, which had been followed up for more than 10 years, and the literature review based on recent scientific articles allowed the elaboration of this text, which met the initial objectives of the research.

DISCUSSION

The Brazilian public health reality does not have in its daily routine the performance of humanized end-of-life processes that allow the patient to choose to interrupt the proposed treatments, especially in patients as young as the one in the reported case. Death processes are still seen as taboo by most of Western society, and therefore, decisions such as the one made by this patient can generate internal conflicts in the care team [6].

These conflicts are moral and ethical, based on religion or even on values passed down by the local culture, but medical practice requires a professional posture, where sometimes it is necessary to abstain from convictions and only follow the course desired by the one to whom the medical service is provided. This will is formalized by the Advance Directives of Will (DAV), which are filled out before the need to make a medical decision, in the present or future, and which manifest the patient's will if he is not able to arbitrate on himself on that occasion [6,7].

This document serves to maintain the patient's autonomy over himself in case he loses his cognitive functions and it is necessary to institute, maintain or suspend therapies that may alter his life prognosis [6]. The case of the reported patient fits into this context, since she is already of legal age, and therefore is responsible for herself. In view of the clinical conditions she carried, the long-term prognosis did not speak in favor of the full recovery of quality of life, which motivated her to give up advanced support in the future.

Although it is not precisely possible to state how long the support associated with instituted treatments would serve to maintain the patient's life, one must consider the psychological suffering to which the patient was being exposed, being away from the mother and other family members during the periods of hospitalization for treatment. Although very young, she had already been living this routine for more than a decade, which compromised part of her childhood and adolescence.

Although the depressive condition was apparent, in view of the need to formalize a SAD, it was necessary to rule out a lupus psychosis. Although considered rare in the literature, if there was such a diagnosis, the patient could not be considered able to understand her health status and decide on the future of her own treatment, which would prevent the elaboration of the document [4,6].

To determine that a patient has full capacities to understand and decide about their health status, 4 basic capacities are taken into account: compression, evaluation, reasoning, and the ability to express their final decision. These indicators show, respectively, whether the patient understands his or her current condition and health prognosis well, whether he or she is able to evaluate the existing alternatives for his or her case, whether he or she is able to compare the outcomes of each possible decision, and whether he or she can formalize his or her will in an understandable manner [6].

Recent studies show that hospitals that make staff and patients aware of AVDs and adopt them in their routine have lower rates of invasive interventions in the last days of the patient's life, including length of stay, admission to intensive care unit (ICU), death at home, CPR maneuvers, use of vasoactive drugs, mechanical ventilation, nutrition, hemodialysis and other invasive procedures in the last days of patients' lives [7]. In the face of unfavorable prospects, it should be considered that ensuring the minimum comfort and quality of life for the patient should be considered a priority.

While opting out of life-sustaining treatments in extreme cases has been shown to improve the end-of-life quality of life for many patients, the positive experience is still confined to a few countries and health centers [7]. Although there is a cultural and moral barrier to be overcome, the idea of dying in comfort needs to be disseminated, so that the implementation of AVDs is broader and allows more patients, when necessary, to have access to this tool for maintaining autonomy.

The tendency is that, little by little, it is understood that the refusal on the part of the patient to continue receiving medical care and support is not an act of rebellion; In some cases, medicine does not have much to offer patients, and they deserve to be fully aware of this, so that they can rationally deliberate about their future and choose how far to insist and when to stop, to enjoy the inevitable last days with those they love, living as close to a normal life and as comfortably as possible.

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