Acute transverse myelitis in systemic lupus erythematosus: report of a case

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Abstract

Systemic lupus erythematosus is an autoimmune disease that usually develops neurological manifestations in a high percentage of the cases. Acute transverse myelitis is a rare neurological complication with significant possibility of damage, sequelae and poor prognosis. We present the case of a patient with systemic lupus erythematosus and acute transverse myelitis who responded adequately to treatment with intravenous steroids and cyclophosphamide. Having in mind acute transverse myelitis as a possibility in any patient with systemic lupus erythematosus, allows us to be ready and able to diagnose and treat this complication early, avoiding sequels and poor prognosis.

Key words: Systemic lupus erythematosus, acute transverse myelitis.

Introduction

Systemic lupus erythematosus (SLE) is an autoimmune disease that can cause multiple organ damage. It is more common in females with a ratio of 9:1 with respect to males. (1)

Neuropsychiatric manifestations in patients with SLE are common, and when these manifestations are developed, the course of the disease and the prognosis are significantly worse. (2)

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The reported prevalence of neuropsychiatric manifestations in SLE is highly variable. There are studies that report prevalence as high as 80-90%. (3) According to latest reports, the prevalence is between 30-40%, and the presentation is possible even before the diagnosis of the disease. (4)

According to the American College of Rheumatology, a total of 19 neuropsychiatric syndromes that can occur in the course of SLE have been described and defined. (5) The most common syndromes are cognitive disorders, mood disorders, headaches, seizures and cerebrovascular disease. (6) The presence of myelopathy in patients with SLE is one of the less common neuropsychiatric manifestations. The reported prevalence varies according to different reports, but it is estimated that occurs in 1-3% of these patients. (7) The aim of the present study is to report a case of transverse myelitis in a patient with SLE and compared it with the previous reports.

Description of clincial case

This is an 18 year-old female, occupation day laborer, with

a past medical history relevant for lupus erythematosus diagnosed 2 years ago in treatment with chloroquine, prednisone, azathioprine, which uses improperly. She was hospitalized 2 years ago for incomplete abortion and left pleural effusion.

His current condition was started 2 months ago with fatigue, weakness, lost of appetite. Progressively she developed a moderate, throbbing type, generalized headache without irradiation and without accompanying symptoms which responded to NSAIDs adequately. Ten days before admission, she started to experience an unquantified rise in temperature with prevalence in the evening, accompanied by nausea and vomiting of gastrobiliar content, diarrhea without mucus or blood, oral intolerance and suddenly in a short period of time (24 hours approximately), she developed a significant reduction of strength and sensitivity of pelvic limbs and loss of sphincter control.

The most relevant findings at her physical examination were: BP 100/60 mmHg, HR 85/min, T 36 ºC, RR 18/min, weight 40 kg, height 149 cm, BMI 18. Patient was conscious, oriented, cranial nerves appeared unaltered. Eye fundus examination was normal. Muscle strength and tendon reflexes: muscle strength 5\5 in upper limbs, biceps, ulnar and triceps reflexes ++. Pelvic limbs with muscle strength 1\5. Patellar and Achilles reflexes +. The Babinski reflex was absent. Superficial and deep sensitivity was abolished in pelvic limbs. There were no other physical examination abnormalities.

The most relevant laboratory abnormalities were the presence of leukocytes in 10,400/mm^3, Hb at 8.8 g/dl with a mean corpuscular volume of 90 and a platelet count in 159,000. Serum electrolytes and liver function test within normal limits. The erythrocyte sedimentation rate was 28 mm/hr and the lumbar puncture showed transparent liquid with a density of 1.010, pH 8.0, total leukocytes 3. No bacteria were seen on Gram stain, glucose 75 mg/dl, protein 130.42 mg/dl, lactic dehydrogenase 91 IU/L. Nerve conduction tests and electromyography showed severe myopathic damage.

The thoraco-lumbar MRI in T2 sagittal sequence showed a hyperintense lesion of irregular morphology, covering more than 2 vertebral bodies, heterogeneous, with uptake after injection of contrast, compatible with acute transverse myelitis (**Figure 1**). The patient was treated with intravenous steroids and cyclophosphamide with favorable results.

Discussion

Acute transverse myelitis is a complication which occurs following an inflammatory process associated with many etiologies. The prevalence in the general population has been reported in up to 4 or fewer cases per million people per year. (8)

Acute transverse myelitis is not a complication associated only with SLE. It can occur in association with many diseases, including sarcoidosis, multiple sclerosis and a variety of connective tissue disorders. It can be a paraneoplastic syndrome and it has been reported in association with infectious diseases such as syphilis, tuberculosis, AIDS, herpes, Mycoplasma pneumoniae, among others. It has been estimated recently that up to 10% of cases are cases of idiopathic acute transverse myelitis. (9)

Among the diseases of the connective tissue, SLE is probably the most associated with acute transverse myelitis. It has been reported also in association with antiphospholipid antibody syndrome and there are reports with other connective tissue diseases very sporadic. (10) It is not known with certainty the mechanism by which SLE can lead to affect the spinal cord in acute transverse myelitis. There are some theories suggesting that the damage is through the formation of arterial thrombi and vasculitis at the level of the spinal cord. (11) An interesting fact is that patients with acute transverse myelitis usually present with other neurological symptoms of SLE, such as depression, seizures, psychosis, among others. There are even reports of acute transverse myelitis preceded by aseptic meningitis (12) and a significant percentage of patients are associated with optic neuritis, which is known as Devic's syndrome. (13)

In the case of our patient, the neurological symptom reported was headache and there were no visual disturbances at any time in the course of the disease.

This complication usually affects young women and is an expression that occurs early in the diagnosis of SLE, as in the case of our patient. There are even

a high percentage of patients who present with this syndrome as the first manifestation of SLE, and nearly half of patients have a recurrence of myelitis. (14) The clinical manifestations are varied, but usually start with numbness of lower extremities with decreased sensation and muscle strength bilaterally to evolve into a great majority of cases to paraplegia. All these symptoms occur in a matter days or even hours. (7)

The presentation of other symptoms before and during the episode of myelitis is common, such as malaise, photophobia, dizziness, neck stiffness, fever, urinary retention and fecal incontinence. (12) There are reports of catastrophic presentations of transverse myelitis with important involvement of both spinal cord and brain tissue at the level of the brainstem, associated with respiratory depression and poor prognosis. (15) The imaging study of choice for diagnosis of acute transverse myelitis is magnetic resonance imaging (MRI). It is common to find spinal cord edema at the site of the lesion and central enhancement of the signal in the same place at level of T2. It is reported that up to 30% of patients do not present a normal image of the spinal cord in MRI. (16) Another important point is to mention the findings in the cerebrospinal fluid (CSF). It is common to observe an increase of polymorphonuclear cells, elevated protein, hypoglycorrhachia and there are some reports about detection of the characteristic antibodies of SLE and probable evidence of intrathecal production of these antibodies. (17) There are no well established recommendations regarding the treatment of choice for these patients. The use of steroids and cyclophosphamide is recommended as quickly as possible during the presentation of this complication because the best prognosis has been obtained from earlier therapy. (7)

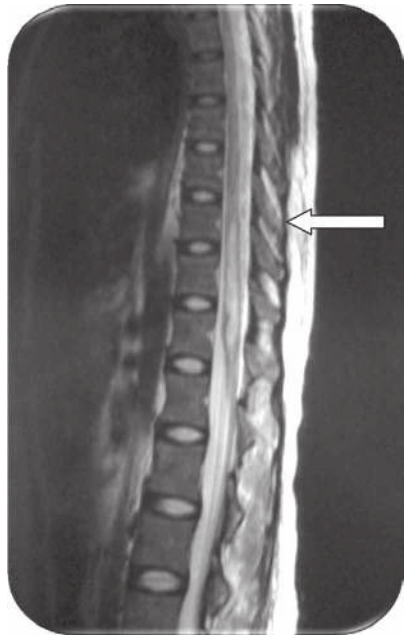
There are reports of cases where the disease is refractory or recurrent and plasmapheresis or intravenous immunoglobulins have been used with controversial results. (18) It is not well established yet, but some recommend the use of antithrombotic therapy in patients with transverse myelitis and positive antiphospholipid antibodies. (10) There are reports in which rituximab has been used with apparent success. (19) On the other hand, there have been cases in where using other drugs such as mycophenolate mofetil with dexamethasone (cases of myelitis persistent and resistant to treatment) with almost complete relief. (20) Intrathecal therapy with methotrexate and dexamethasone have been used successfully, (21) and interestingly there are reports in where bone marrow transplantation has been used, specially for recurrent and refractory cases with apparent success. (22)

Comments and conclusions

Among the many complications that can occur in patients with SLE, it is always important to consider the neurological manifestations due to their high prevalence. Some of these complications such as acute transverse myelitis may not be very common, but regained its importance due to damage, disability and the potential consequences that may occur.

Having in mind acute transverse myelitis as a possibility in any patient with SLE, especially in the early diagnosis, allows us to be ready and able to diagnose and timely treat this complication early and avoid important sequels and poor prognosis.

Figure 1. Thoracolumbar MRI T2 sagittal sequence



Legend: It is observed an hyperintense lesion with irregular morphology that spans more than two vertebral bodies on T2 sequence, heterogeneous, and shows moderate uptake after injection of gadolinium, compatible with acute transverse myelitis.

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