A case of neuropsychiatric lupus with myelopathy successfully treated with corticosteroids

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Summary

This report describes a 16-year old female patient who presented with acute paresis in both lower limbs, acute urinary retention , blurred vision and arthritis. The patient was diagnosed as having systemic lupus erythematosus with myelitis and bilateral abducent nerves palsy. The administration of steroids resulted in marked improvement in her neurological symptoms.

Key words: ***Systemic Lupus Erythematosus, Neuropsychiatric, Myelopathy, Corticosteroids.***

Résumé

Ce rapport tâche de décrire une patiente âgée de 16 ans qui s'est presentée atteinte du paresis chronique dans les deux membres inférieurs, retention de l'urine chronique, vue trouble, et l'arthrite. La patiente a été diagnostique atteinte du lupus erythemateu systemique avec la myelite et la paralysie abducent des nerfs bilateraux, (bilateral abducent nerves palsy). L'administration des stériodes a provoqué une amélioration remarquable dans ses symptôms neurologiques.

Introduction

Systemic lupus erythematosus (SLE) is a chronic multisystem autoimmune disorder of unknown cause affecting mainly women of childbearing age [1] . SLE can affect the skin, joints, kidneys, lungs, nervous system, serous membranes and/or other organs of the body. Distinct immunologic abnormalities, especially the production of a number of antinuclear antibodies, are other prominent features of the disease. The clinical course of SLE is characterizeu by periods of remissions and chronic or acute relapses. Treatment is based on preventive measures, reversal of inflammation, prevention of organ impairment, and alleviation of symptoms[2-4].

Central nervous system (CNS) involvement has been reported to occur in 14 to 75 % of patients with SLE [5-6]. Myelopathy is a well recognized, but infrequent and serious neurological manifestation of systemic lupus erythematosus, occurring in less than 1% of SLE patients [7].

In this case report we demonstrate the clinical features in a 16 year old Saudi female who presented with systemic lupus erythematosus with myelopathy, peripheral neuropathy and cranial nerve lesion (bilateral abducent nerves), successfully treated with pulse and oral corticosteroid therapy.

Case report

A 16-year-old Saudi girl presented to the Emergency Room of Assir Central hospital, Abha, Saudi Arabia on 9 July 2002 with a sudden attack of blurred vision, retention of urine and inability to walk. Her condition started 2 years ago with recurrent attacks of mild unilateral headache manage successfully with analgesics in the local primary health center as a case of migraine Two months prior to admission, her headache progressed in severity, became continuous, generalized, and was associated with non-projectile vomiting. In the preceding one month, she started having blurring of vision and double vision which occurred when looking extremely to the left or right side. The patient's relatives also noticed squinting of her left eye. At this time, she started having unusual hair fall, asymmetrical joint pain and swelling involving small joints of the hands and feet, elbows, knees and ankles. For the last 25 days she has had a rapidly progressive low back pain, not radiating or related to cough or straining but associated with paraesthesia of both lower limbs. In the 24 hours preceding her admission the patient had retention of urine with marked weakness in her lower limbs

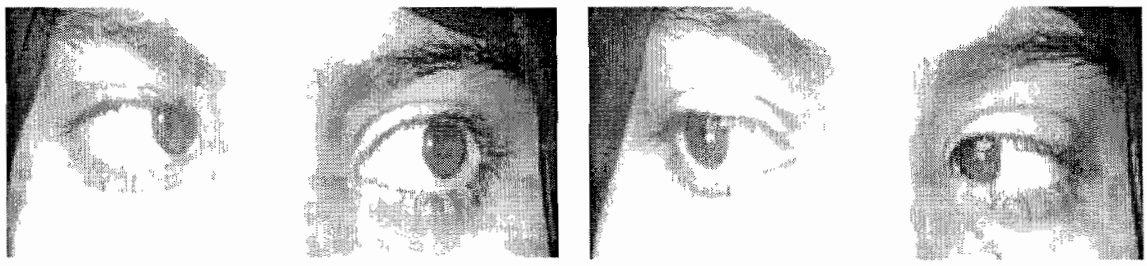

Fig. 1 ***Partial abducent nerve palsy. The left side is more involved than the right side***

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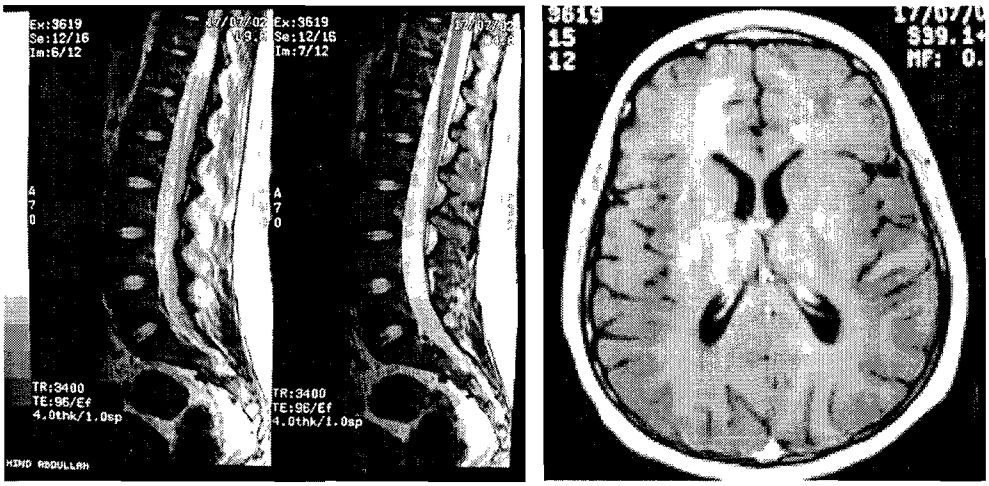

Fig. 2 *MRI of the lumbo-sacral spine (A) and the brain (B). No lesions are evident.*

associated with inability to walk. On examination, she was conscious, well oriented, looked ill, was in pain, but was neither pale, cyanosed nor jaundiced. She was not in respiratory distress. There were no palpable peripheral lymph nodes, no skin rash, no digital clubbing. There was no lower limb edema and the jugular vein pressure was not raised. Her vital signs were normal BP= 110/70/mmHg, Heart rate 72 beat/minute) . Chest, cardiovascular and abdominal examinations revealed no abnormalities. Right knee and left ankle effusion were present. Central nervous system showed bilateral abducent nerve lesions (Fig.1). There was hypotonia involving both lower limbs with decreased power bilaterally (right side (1-2/5) more than the left side (4/5) in all muscle groups) . Knee and ankle reflexes were brisk bilaterally. Plantar reflex was flexor on the left side and extensor on the right side. Sensation was diminished in both feet.

Investigations. The ESR (112mmhg) was elevated, there was normocytic, hypochromic anemia (Hb=8.6 gm/dl), lymphopenia, and mild proteinuria (280mg/24hours) with normal kidney function. Antinuclear antibody (ANA) and anti double stranded deoxyribonuclic acid (dsDNA) were positive, but cytoplasmic antineutrophil cytoplasmic antibody (cANCA) and perinuclear antineutrophil cytoplasmic antibody (pANCA) were negative. Serum complement C4 and C3 were low (C4 = 6.44mg/dl, C3 = 24.4mg/dl) . The spinal fluid analysis was normal. Plain chest radiography, plain X-ray of the spine and abdominal ultrasound scan were all normal. Magnetic Resonant Imaging (MRI) of the brain and spine were within normal limits (Fig.2). Electromyography (EMG) and nerve conduction studies showed a mixed picture of peripheral neuropathy and myopathy. Treatment was started with pulse methylprednisolone therapy (1 gm / intravenous) for three successive days followed by daily oral prednisolone (1mg/Kg) for another one month. The patient showed progressive neurological and arthritic improvement with gradual decreasing in her ESR (46mmHg on discharge) and was discharged on 18 August 2003 with adequate power in her lower limbs (power 4/5), residual abducent dysfunction. She is continued on oral corticosteroid (40 mg/day) and physiotherapy program.

Discussion

Systemic lupus erythematosus (SLE) has been known for almost a century and remains the prototypic immune disease [8]. SLE predominantly affects women and is more common in blacks[8]. Although survival rates have improved, over one-half of patients with systemic lupus erythematosus have permanent damage in one or more organ systems. Arthritis and cutaneous manifestations are the most common presentations, but renal, haematologic and neurologic manifestations contribute largely to mortality and morbidity [8].

Central nervous system involvement in SLE is frequent and severe[9]. However, myelopathy is rarely reported[9],occurring most often during the course of the disease [10]. Although magnetic resonance imaging (MRI) is the modality of choice for diagnosis of myelopathy. It shows signal abnormalties, usually T2 hypersinensity, focal or extensive gadolinium enhancement and sometimes cord swellings. However about 40 % of acute transverse myelopathies remain undemonstrated [11]. In our case MRI of the spine and brain were normal. Zenone et al.,[10] reported that MRI findings depend on the timing of the examination and the stage of the disease, the MRI may therefore be normal [11].

The treatment of patient with neuropsychiatric lupus can be difficult and complex owing to the variety of nervous system manifestations that can occur [12]. Also, the best treatment protocol has not yet been agreed on[10]. Currently, there is controversy about cycolophosphamide and methylprednisolone for the treatment of neuropsychiatric manifestations. In the patient presented, treatment with pulse methylprednisolone followed by a high dose of oral corticosteroids produced marked clinical improvement.

Although there are many recommendations in the

literature suggesting aggressive therapy with steroids and cyclophosphamide to get best outcome, Trevisani et al.,[13] concluded in their review that cyclophosphamide treatment regimen in neuropsychiatric involvement in SLE has no evidence to show superior effectiveness and safety when compared with methylprednisolone [13].

Conclusion

In the case presented, early treatment with high intravenous doses of corticosteroids, was effective and produced a good outcome in this SLE patients with neuropsychiatric manifestations.

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