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Case Report

Transverse myelitis in mixed connective tissue disease

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SUMMARY *A case of transverse myelitis in a 16-year-old woman with mixed connective tissue disease is described. After treatment with azathioprine and prednisolone complete recovery was obtained.*

Key words: Mixed Connective Tissue Disease, Transverse Myelitis.

INTRODUCTION

Mixed connective tissue disease (MCTD) is a disease for which there is no formal criteria for classification. The diagnosis requires the presence of antibodies to ribonucleoprotein (anti-RNP). Common clinical signs are arthritis, swollen fingers, Raynaud's phenomenon, myositis and pulmonitis. Neurologic disease is commonly regarded as rare and most often manifested by trigeminal neuralgia (1). We report, to our knowledge, the first case of recovery after transverse myelitis in MCTD.

CASE REPORT

A 16-year-old woman with a two-year history of MCTD manifested by arthralgia, alopecia, swollen hands, Raynaud's phenomenon, myositis, positive test for antinuclear antibodies, high titer of anti-RNP and negative test for anti-DNA was admitted to hospital because of progressing weakness of all four extremities. One week prior to admission prednisolone was increased from 10 to 30 mg/daily since active myositis was suspected.

On physical examination the patient appeared acutely ill. The temperature was 38.2°C, pulse 80/min and blood pressure 130/80 mm Hg. Alopecia and vasculitis of the palms of both hands were present. Auscultation of heart and lungs was normal. On abdominal examination the bladder was recognized above the symphysis and urinary retention was confirmed by catherization. All four extremities were weak, the lower extremities being most severely affected. Tendon reflexes were absent on both upper and lower extremities. Bilateral extensive plantar responses were found. In the following 48 hours the paresis of the lower extremities progressed, rectal sphincter tone became weak and a sensory level below Th 12 were found.

Laboratory examination included: hemoglobin 6.6 mmol/l, total white blood cell count 4.9 $\times$ 10^9/l, lymphocytes 0.3 $\times$ 10^9/l, platelet count 135 $\times$ 10^9/l, Wester-

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gren sedimentation reaction 49 arb units/hour. Creatinin, urea and creatinin-phosphokinase were normal. Initially urinalysis showed pyuria and hematuria but became normal after treatment for a urinary tract infection. The anti-RNP titer exceeded 204,800. Antibodies to DNA were present in small amounts, 0.41 arb units (normal range 0.00-0.30 arb units). Spinal fluid examination done after initiation of therapy indicated: glucose 2.3 mmol/l, protein 1.7 g/l and albumin 14.5 micromol/l. Cells were absent. A brain-computerized tomography scan revealed slight cortical atrophy, but was otherwise normal. Neurophysiological study demonstrated myopathia (reduced amplitude on maximal muscle contraction). Conduction velocity in peripheral nerves was normal. Urodynamic examination revealed neurogenic bladder dysfunction. Two days after admission treatment with azathioprin 100 mg/daily was started and prednisolone was increased to 60 mg/daily. The patient improved and after four weeks of treatment muscle function was normal. After a further four weeks of treatment control of bladder function was regained. Neurological examination was normal. The course was complicated by several urinary tract infections and a sacral decubitus, which needed transplantation. Prednisolone was slowly tapered and azathioprin was stopped after six months of treatment. At follow-up 20 months after onset of myelitis, the patient was well, apart from a tendency to urge incontinence. Symptoms of myositis were controlled by prednisolone 10 mg/daily.

DISCUSSION

Classification of MCTD can be problematic since pathognomamic features are lacking. Our patient had, however, clinical signs which are considered characteristic of MCTD: swollen hands, arthralgia and Raynaud's phenomenon. Furthermore, she had very high titer of anti-RNP. Although anti-DNA was briefly present in the course of the disease, the patient did not meet the 1982 revised criteria for the classifiçation of systemic lupus erythematosus (SLE) (2) since abnormal microurine was present only when the patient had urinary tract infections. Our patient had a long history of myositis; although serum creatininekinase level was normal, it cannot be excluded that recent activity in the myositis may have contributed to the muscle weakness observed. However, the patient had clinical signs of spinal cord damage as well, paraplegia, sensory loss and loss of sphincter control. There were no signs of other diseases which could cause spinal cord damage (syphilis, multiple sclerosis or vitamin B_{12}-deficiency), and polyneuropathia was excluded by EMG. We believe the patient suffered from transverse myelitis; this was supported by the characteristic finding of a high protein content and a low glucose content in the spinal fluid.

Transverse myelitis is a rare complication in MCTD and so far only a single case has been reported (3). This patient had the serologic changes seen in MCTD. Clinical features characteristic of MCTD, however, were absent, and the patient met the criteria for classification of SLE. Thirty-six cases of transverse myelitis have been reported in SLE (4,5), and a review of these cases are given by Al-Husaini and Jamal (5). The characteristic neurologic findings are paraplegia, sensory loss and loss of sphincter control. Spinal fluid examination often shows high protein and low glucose content. The treatment of transverse myelitis has been unsuccessful since only few patients have recovered completely. The effect of glucocorticoids is debated, but seems to modify the disease favourably if given early after the onset of myelitis (4,5). Recently Warren and Kredish (6) described a case in which the patient recovered quickly and completely, following treatment with intravenously methylprednisolone 1 g daily. Thus, transverse myelitis is a serious complication of MCTD or SLE. Treatment with glucocor-

ticoids or a combination of glucocorticoids and cytostatic drugs may prove useful if given in the early stage of the disease.

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Book Review

Rhumatologie - Pathologie osseuse et articulaire.
A. RYCKEWAERT, Médecine-Sciences Flammarion, Paris, 1987, 492 pp., 300 figures, 495 FF.

This is a remarkable book. The author, a distinguished French professor in rheumatology, should be congratulated for the original presentation and upgrading of the developments over the past 15 years. This is one of the best books I have ever consulted on rheumatology over the last 20 years. This book treats the broad field of medical osteology and rheumatology concisely on the one hand and exhoustively on the other. The 300 drawings which illustrate the bone lesions, make this book particularly didactic. The reference list after each chapter is updated and easy to consult because the references are listed for each disease and syndrome separately. It is a pity that this book is written in French, because many more rheumatologists, students, radiologists, orthopaedic surgeons and allied professionals in rheumatology and medical bone diseases over the world could gain profit and consult it every day if it were written in English.

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