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Case report

Epilepsia partialis continua in a patient with Behçet's disease☆

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Abstract

Behçet's disease (BD) is a multisystemic, recurrent, inflammatory disorder of unknown aetiology. Neurological involvement is characterised either by primary parenchymal lesions or secondary to major vascular involvement. Seizures are rarely seen in BD and their occurrence can be related to seizure provoking factors or exacerbation of the disease. We experienced a case of neuro-BD presenting with subacutely developing mental and behavioral changes, followed by left dominant tetraparesis with bilateral pyramidal signs, fever and left hand focal motor seizures with elementery clonic motor signs which later evolved into right hand epilepsia partialis continua (EPC) of Kojevnikov. The seizures were very resistant to antiepileptic drugs and 8 months after neurological involvement the patient died. The EPC evolving after neurological involvement is associated with high mortality rate.

Keywords: Epilepsia partialis continua; Seizure; Neurological involvement; Behçet's disease

1. Introduction

Behçet's disease (BD) is a heterogeneous, multisystem, recurrent, inflammatory disorder of unknown aetiology [1]. Central nervous system (CNS) involvement is one of the most serious manifestations of BD and is seen approximately 4–49% of the patients with BD [2,3]. The clinical course of neuro-BD is relapsing–remitting, primary progressive or secondary progressive. Patients develop subacute episodes of neurological dysfunction most commonly situated either within the brainstem or in some cases with hemisphere involvement with mental changes. Seizures are rarely seen in BD, and their occurrence can be related to either seizure provoking factors or exacerbation of the disease itself [2,4,5].

We experienced a case of neuro-BD presenting with subacutely developing mental and behavioral changes, followed by left dominant tetraparesis with bilateral pyramidal signs, fever and left hand focal motor seizures with elementery clonic motor signs which later evolved into right hand epilepsia partialis continua (EPC) of Kojevnikov. The seizures were classified according to new proposed criteria of the International League Against Epilepsy [6]. To our knowledge, EPC of Kojevnikov has not previously been reported in neuro-BD and its occurrence would appear to be associated with high mortality.

2. Case report

A 37-year-old woman was referred to us from a state hospital with fever, lethargy, affective and behavioral changes as well as left hemiparesis. In her past medical history she had suffered recurrent oral and genital ulcerations since the age of 20 and in addition she had had bronchial asthma for 2 years. She had not been a diagnosed with BD but had received symptomatic treatment for pulmonary disease during the last 2 years. Her sister has BD and her mother has also recurrent oral and genital ulcerations but not diagnosed with BD either. In the state hospital she received i.v. methyl pred-

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nisolone treatment 1 g/day for 3 consecutive days followed by a cyclophosphamide regimen, but did not recover. As a result she was referred to our department. On examination, her blood pressure was 120/80 mmHg, pulse was 80/min, fever was 38 °C. The general physical examination was consistent with multiple genital scars, erythema nodosum-like lesions on the lower extremities and scattered papulopustular hyperactive lesions on all extremities. A neurological examination revealed coma vigil (apparent vigilance in a state of imperceptiveness and unresponsiveness), facial diplegia, absent pharyngeal reflexes, left prominent tetraparesia with bilateral pyramidal signs, and left hand and arm focal motor seizures with elementery clonic motor signs suggesting widespread CNS involvement.

Biochemistry, serum electrophoresis, folate, B12 and immunglobulins were all normal, haematology showed a moderately increased erythrocyte sedimentation rate of 55 mm. Rheumatoid factor (RF) titre, syphilis and brucella serology, HIV antibodies were negative however CRP levels were high. Coagulation studies showed a positive antiphospholipid antibody level. In addition, HLA B5 was also positive. CSF pressure was normal but the analysis was abnormal with 33 lymphocytes, protein was 29.8 mg/day, glucose was 51 mmol/l (serum 107 mmol/l). CSF cytology, chest radiography, ECG, transthoracic echography were all normal. However, the interictal EEGs showed severe encephalopathy with generalised theta and delta waves but no epileptiform features. A brain MRI was performed on three occasions. On the first MRI examination, multiple lesions were seen in the cerebral peduncles, subthalamic region and right frontal lobe. The lesions were hypointense on T1-weighted images and hyperintense on T2-weighted and fluid-attenuated inversion recovery (FLAIR) images. Only the lesions in the right frontal lobe showed mild contrast enhancement (Fig. 1). There was no evidence of venous thrombosis.

The diagnosis of Behçet's syndrome with neurological complications was made on the basis of recurrent oral and genital ulcerations, skin lesions, HLA B5 positivity and multiple focal CNS lesions [7]. Intravenous methyl prednisolone therapy 1 g/day for 5 consecutive days was administered followed by 0.4 g/kg/5 day IVIG treatment. Following this her vigilance slightly improved but otherwise no prominent changes were observed so prednisolone 0.5 mg/kg/day was administered. Additionally, carbamazepine therapy at a dose of 400 mg/day was administered for her left arm focal motor seizures however no response was observed so the medication was modified to 1500 mg Na–valproate and 200 mg lamotrigine regimen. Consecuently, the frequency and duration of the seizures were decreased but were never completely controlled. In the follow-up period, the patient suffered deep venous thrombosis and urinary tract infection, both of which were appropriately treated with antibiotics and anticoagulants.

One month following admission the seizures were still rarely apparent on the left arm. Then, after a short period her right hand and arm began to be affected by focal motor seizures with elementery clonic motor signs as well. Gradually these evolved into epilepsia partialis continua (EPC) of Kojevnikov. Apart from this her neurological state remained the same. The ictal EEG showed generalized slowing with delta and theta waves and T5 maximum rhythmic spike activity on the left frontotemporal region (Fig. 2). The second MRI was performed during the right arm EPC. The lesions in the cerebral peduncles and subthalamic region had completely disappeared and the lesion in the right frontal lobe had decreased markedly in size (Fig. 3). After the patient was moved to the intensive care unit (ICU) and put on general anesthesia with penthobarbital, her antiepileptic drugs (AEDs) were re-evaluated and modified to 3000 mg/day Na–valproate, 200 mg/day lamotrigine, 300 mg/day phenobarbital and 2400 mg/day gabapentin regimen. She remained under observation in the ICU for 7 days, during which time her seizures reduced in frequency but never completely stopped.

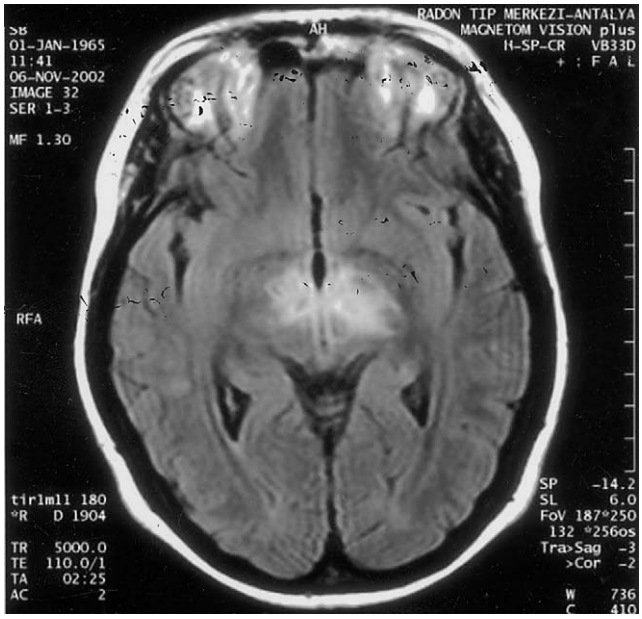

Fig. 1. Fluid-attenuated inversion recovery (FLAIR) weighted transverse MR image shows hyperintense lesions in cerebral peduncles and subthalamic region.

Three months after of her admittance the patient's neurological picture had not changed markedly though she suffered a series of hospital infections. Her left hand focal motor seizures with elementery clonic motor signs were almost completely controlled with AEDs although her right hand focal motor seizures were still being observed particularly after sensory stimuli; such as sudden noise or touch. Apart from this there was no change in her neurological picture. The third MRI, performed the fifth month of her admittance, did not show any difference from the second MRI. She died eight months after the neurological involvement as a result of septicemia.