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Epileptic seizures in Neuro-Behcet disease: Why some patients develop seizure and others not?

Gulnihal Kutlu [a,*], Sencer Semercioglu [b], Serap Ucler [b], Abidin Erdal [b], Levent E. Inan [b]

[a] Mugla Sitki Kocman University, School of Medicine, Department of Neurology, Mugla, Turkey
[b] Ministry of Health Ankara Research and Training Hospital, Department of Neurology, Ankara, Turkey

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ABSTRACT

Purpose: Behcet disease (BD) is a chronic relapsing inflammatory disorder. Neuro BD (NBD) is seen in approximately 5% of all patients. The aim of this study is to investigate the frequency, type and prognosis of epileptic seizures in different forms of NBD.
Methods: All files of 42 patients with NBD were evaluated between 2006 and 2012, retrospectively. The demographic data, the presentation of NBD, clinical findings including seizures, EEG and neuroimaging findings were reviewed.
Results: The mean age of patients was 35.02 ± 8.43 years. Thirty (71.4%) patients were male; the remaining 12 of them were female. Twenty-four patients had brainstem lesions; 16 patients had cerebral venous thrombosis. Spinal cord involvement was seen in two patients. Seven patients had epileptic seizures (six partial onset seizures with or without secondary generalization). Six of them had cerebral sinus thrombosis (CVT). Four patients had a seizure as the first symptom of the thrombosis. One patient had late onset seizure due to chronic venous infarct. The other patient with seizure had brainstem involvement. The remaining was diagnosed as epilepsy before the determination of NBD.
Conclusion: CVT seen in BD seems to be the main risk factor for epileptic seizures in patients with NBD. The prognosis is usually good especially in patients with CVT. Epileptic seizures in patients with brainstem involvement may be an indicator for poor prognosis. Superior sagittal thrombosis or cortical infarct would be predictor of seizures occurrence because of the high ratio in patients with seizures.

1. Introduction

Behcet's disease (BD) is a chronic relapsing inflammatory disease of unknown etiopathogenesis [1–3]. It is a disorder of young adults with male predominance. There is a geographical variation in prevalence [4]. Hulusi Behcet, Turkish dermatologist, first described it [2]. BD is mainly characterized by oral and genital ulcerations, ocular manifestations and by involvement of the gastrointestinal, cardiovascular, pulmonary, skeletal and central nervous system [1]. Recurrent oral ulceration is prerequisite with any two of genital ulcerations, skin lesions, uveitis and hyperactivity of skin to nonspecific physical insult (Patergy test) according to the diagnostic criteria formed by the International Study Group [5].

Neurologic manifestation in BD (NBD) is involved in about 5–10% of the patients with BD [3,6]. There are two major forms of neurologic involvement of BD according to the clinical and imaging examination. The first, which is seen in the majority of patients, may be characterized as a vascular-inflammatory central nervous system (CNS) disease with focal or multifocal parenchymal involvement, mostly presenting with a brainstem syndrome (intra-axial NBD); the other, which has few symptoms and a better neurologic prognosis, may be caused by isolated cerebral venous sinus thrombosis and intracranial hypertension (extra-axial NBD), occurring in up to 20% of the cases. Spinal cord involvement, arterial CNS involvement, optic neuritis, aseptic meningitis, and peripheral neuropathies may be seen as a presentation of NBD, but are rare [3,7].

Epileptic seizure in NBD was observed rarely in previous studies [8,9] Different seizures types such as partial seizures, generalized tonic clonic seizures; myoclonic jerks, epilepsia partialis continua and status epilepticus were reported [8–12]. The aim of this study is to investigate the frequency, type and prognosis of epileptic seizures in different forms of NBD.

* Corresponding author at: Mugla Sitki Kocman Universitesi Tip Fakultesi, Noroloji AD, Mentese, Mugla, Turkey. Tel.: +90 252 2115180; fax: +90 252 2111345; mobile: +90 532 4173376.
E-mail address: gulnihalkutlu@yahoo.com (G. Kutlu).

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2. Methods

All files of 42 patients with NBD were reviewed retrospectively in Ankara Research and Training Hospital, Department of Neurology between January 2006 and December 2012, retrospectively. The demographic data, the presentation of NBD, clinical findings including seizures, cerebrospinal fluid (CSF) examination, electroencephalography (EEG) and neuroimaging finding were reviewed. Seizures were classified according to the criteria of International League Against Epilepsy [13]. The diagnosis of NBD was revised according to the International Consensus Recommendation (ICR) Criteria for NBD during the retrospective analysis [14]. The clinical course of epileptic seizures and NBD were also evaluated.

3. Results

The mean age of patients was 35.02 ± 8.43 years. Thirty (71.4%) patients were male; the remaining twelve (28.6%) of them were female. All of the patients had definite NBD according to the ICR criteria. Twenty-four (57.1%) patients with NBD had CNS parenchymal involvement with brainstem-diencephalic regions. CVT was present in sixteen (38.1%) while two patients had only spinal cord involvement (5.8%).

Seizures were observed in seven (16.7%) of 42 patients. Demographic data, clinical features, seizure type, CSF findings, EEG, magnetic resonance imaging findings and prognosis are shown in Table 1. The mean age was 30.43 ± 9.36 years (age range 24–49). Only one patient with seizure was female, the remaining six (85.7%) patients were male. Mean age at the diagnosis of BD 27.0 ± 8.68 (16–42 years) years. The patients with epileptic seizures were followed up between 3 months and 5.5 years (mean: 32.14 ± 22.37 months). In their medical history, six patients had no risk factors regarding the epilepsy; only one patient had the family history for epilepsy. The mean duration of NBD in patients without seizures was 30.34 ± 22.24 months.

CVT was observed in six patients with epileptic seizure. Cranial magnetic resonance imaging and venography was performed to all patients. Superior sagittal thrombosis was present in four patients (66.7%) while two patients had left transverse sinus thrombosis (Fig. 1A and B). However, the ratio of superior sagittal thrombosis was 40% when the patients diagnosed as CVT without seizures were analyzed.

Among the patients with CVT, five of them had complex partial seizures and three of them had also secondary generalized tonic clonic seizures (SGTCS). Four patients had seizures as the first symptom of the thrombosis. One patient had late onset seizure probably due to chronic venous infarction in left temporal–parietal region seven weeks after the diagnosis of NBD disease. The seizures of other patient were started three months before the determination of thrombosis.

Patient 1 and 2 had only one partial onset and SGTCS seizure at admission. They were treated with carbamazepine and levetiracetam, respectively and antiepileptic drugs were stopped 6 and 8 months of treatment. No seizure was seen in their follow up. Their EEGs were also normal. Patient 3 had two complex partial seizures, whereas three partial onset and SGTCS was present in Patient 4. They were treated oxcarbazepine and carbamazepine during 24 months, respectively. The dose of antiepileptic drug was decreased in Patient 3 and 4 and termination of treatment is planned for them. 4–7 Hz slow waves were seen in their first EEG and recurrent EEG examinations were normal. Seizure was not recurred in both of patients. Patient 5 (only female patient) had venous infarction and she was admitted to the hospital with complex partial status epilepticus seven weeks after the diagnosis of CVT. Left temporal seizure activity was also observed in her EEG. Firstly, phenytoin infusion was applied to her and then phenytoin therapy was continued. She was followed up during 66 months after the seizure. The treatment was changed from phenytoin to oxcarbazepine, because she was planning pregnancy. No seizure was occurred before, during and after pregnancy. We decided to terminate the treatment after 52 months from the seizure beginning and decreased the dosage. However, complex partial seizure was started again and the dosage was increased. Recurrent EEG also showed us left temporal sharp waves (Fig. 2). Myoclonic and one generalized tonic clonic seizure (GTCS) were present in

Table 1
Demographic data, clinical features, seizure type, CSF findings, EEG, magnetic resonance imaging findings and prognosis of patients.

No	Age year	Sex	AgeDx BD year	NBD type	Relation NBD	Seizure type	CSF	Clinical manifestation	EEG	MRI MRV	Follow up month	Outcome
1	25	M	24	CVT	Attack	Partial onset SGTCS	–	Headache Papilledema	1.N 2. N	SSS Thrombosis	35	Seizure free AED stopped
2	37	M	35	CVT	Attack	Partial onset SGTCS	–	Headache Papilledema	1.N 2. N	SSS Thrombosis	12	Seizure free AED stopped
3	24	M	22	CVT	Attack	Partial onset	–	Headache Papilledema	1. Slow waves 2. N 3.N	SSS Thrombosis	24	Seizure free AED continue Termination plan
4	28	M	25	CVT	Attack	Partial onset SGTCS	–	Headache Papilledema	1. Slow waves 2. N	TS Thrombosis	30	Seizure free AED continue termination plan
5	26	F	25	CVT	Course	Partial onset SE	High Pro Pro: 68 mg/dl Mild Pleocytosis	RHH Papilledema	1. Seizure act 2. L T Sharp w 3.LTSharpw 4. L T Sharp w	TS Thrombosis Venous Infarct	66	Seizure free Recurrence when AED decrease Seizure free with treatment
6	24	M	16	CVT	Before Attack	Myoclonic GTCS	–	Headache	1. Spike wave 2. Spike wave 3. Spike Wave	SSS Thrombosis	55	Rare myocloni AED continue
7	49	M	42	Brain stem lesion	Course	Partial onset	High pro Pro: 89 mg/dl Moderate Pleocytosis	Hemiparesis decrease consciousness	1. Slow waves	Brain Stem lesion	3	Seizure free Died

BD, Behcet disease; NBD, Neuro-Behcet disease; CSF, cerebrospinal fluid; CVT, cerebral sinus thrombosis; SGTCS, secondary generalized tonic clonic seizure; GTCS, generalized tonic clonic seizure; act, activity; SSS, superior sagittal sinus; TS, transverse sinus; EEG, electroencephalography; MRI, magnetic resonance imaging; MRV, magnetic resonance venography; HH, hemi-hypoesthesia; AED, antiepileptic drug; JME, juvenile myoclonic epilepsy; Dx, diagnosis; SE, status epilepticus; Pro, protein.