EEG-IMAGING CORRELATION OF EPILEPSIA PARTIALIS CONTINUA IN A PATIENT WITH POSSIBLE NEUROSARCOIDOSIS

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Abstract
Case presentation of a possible neurosarcoidosis with epilepsia partialis continua as the first manifestation. The interesting part of this case is the clinical-anatomical correlation of epilepsia partialis continua as demonstrated by EEG and MRI findings.

Keywords: Epilepsia partialis continua, anterior central sulcus, EEG, neurosarcoidosis, brain MRI

Introduction

Epilepsia partialis continua (EPC) is a rather uncommon condition with a prevalence rate of less than one per million[1]. It is characterized by regular or irregular clonic muscular twitches affecting a limited part of the body, occurring for a minimum of 1 hour and recurring at intervals of no more than 10 sec. A wide range of pathology has been reported to cause EPC.

Sarcoidosis is an inflammatory multisystem disorder that can affect any part of the nervous system. The prevalence of clinical involvement of the nervous system is estimated to be about 5-15%. However isolated neurosarcoidosis is considered to be extremely rare, though its exact prevalence has not be determined[2]. Cranial nerves, hypothalamus, and pituitary gland are the most commonly regions involved, but meninges, parenchyma, brainstem, and spinal cord may also be affected.

Common clinical manifestations are cranial neuropathy, optic neuritis, aseptic meningitis, neuroendocrinological dysfunction, encephalopathy, cognitive dysfunction, seizures, and peripheral neuropathy.

Here we present a rare case of possible neurosarcoidosis with epilepsia partialis continua as the first manifestation.

Case report

A 79-year-old woman was admitted to our hospital with rhythmic clonic jerks of her left upper limb during the past few hours, without loss of consciousness. Her past clinical history was clear except for dyslipidaemia managed with rosuvastatin. On admission, the patient was completely aware of the event and her neurological examination revealed left mild hemiparesis with pyramidal signs.

An ictal EEG was performed, showing focal continuous hemirhythmic epileptiform activity of sharp waves on the central – parasagittal area in phase reversal at C4 electrode -PLEDs on the right hemisphere (Figure 1A). Brain MRI, that was performed three days later, revealed meningeal enhancement of the right frontal lobe, especially of the anterior central sulcus with restricted diffusion on DWI and increased signal on T2 sequence (Figure 2). The above findings were compatible with epilepsia partialis continua.

The routine blood work and further work-up for metabolic, immune-mediated and infectious disorders was within normal limits, apart from a mildly elevated rheumatoid factor (31.2IU/ml). The lumbar puncture revealed 10 leukocytes/μL (lymphocytes), with elevated IgG synthesis (IgG index: 1.09) but with normal protein and glucose. PCR, cytological analysis and cultures of cerebrospinal fluid were negative. Due to elevated serum angiotensin converting enzyme (ACE: 169U/L), a further analysis of CSF was performed, measuring the levels of the enzyme, which was found 3.5 U/L (upper normal limits 2.8).

Chest CT and bronchoscopy were normal. Our patient refused brain biopsy.

Based on the above findings and according to commonly applied criteria, the patient was diagnosed with possible neurosarcoidosis[3]. EPC was controlled within the next twenty-four hours using diazepam, levetiracetam and oxcarbazepine. Additionally, she received a five-day course of intrave-

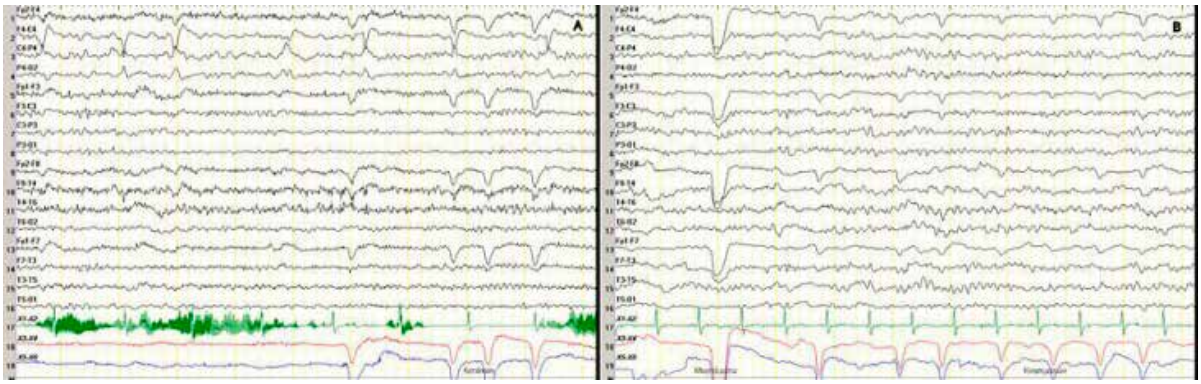

Figure 1. (A) Ictal EEG - Focal continuous hemirhythmic epileptiform activity of sharp waves on the central – parasagittal area in phase reversal at C4 electrode. (B) New EEG 5 months later showing mild diffuse slowness
254x190mm (96 x 96 DPI)

nous methylprednisolone.

Assessment of the patient, 5 months later, revealed clinical and imaging deterioration. She remained seizure – free but the neurological examination revealed a decline in cognitive function. A new MRI scan revealed worsening and extension of meningeal enhancement and parenchymal involvement.

An EEG was performed, showing mild diffuse slowness with paroxysmal focal slow activity on the right frontotemporal region (Figure 1B). Second line treatment with azathioprine was proposed to the patient.

Discussion

Prognosis of EPC depends on age of onset, underling disease and type of EPC, with type I often occurring due to local non-progressive lesion in the sensorimotor cortex and type 2, due to a progressive lesion. Differential diagnosis between progressive and non-progressive encephalopathy is very important in the evaluation of EPC.

Periictal MRI changes at the site of seizure focus with high signal on T2, restricted diffusion and contrast enhancement is a common phenomenon of EPC1. Follow up imaging is required to clarify the nature of MRI findings, i.e. whether these are the results or the causes of the EPC.

In the case of neurosarcoidosis, seizures are estimated to occur in 5–20%. The neurologic manifestations that may contribute to development of seizures are protean and include parenchymal abnormalities including encephalopathy/vasculopathy (5–10%), intraparenchymal mass lesion(s) (5–10%), endocrinopathy with metabolic disturbance including hypothalamic and/or pituitary dysfunction

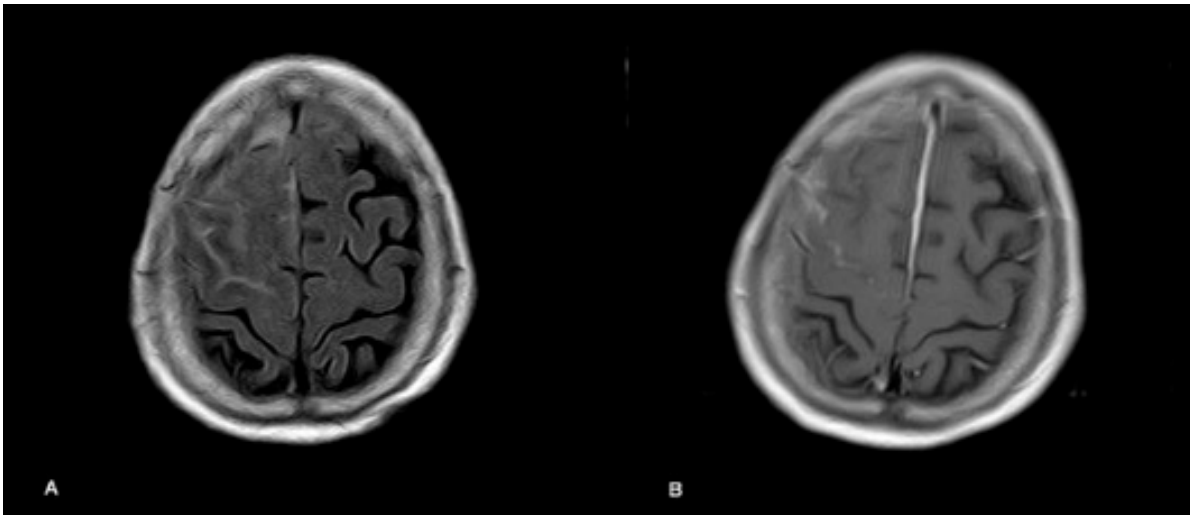

Figure 2. (A) Fluid attenuation inversion recovery sequence showing increased signal in right frontal lobe. (B)T1 C+. Meningeal enhancement of the right frontal lobe and especially of the anterior central sulcus.
254x190mm (96 x 96 DPI)

(10–15%), meningeal disease (including aseptic meningitis and meningeal mass) (10– 20%), and hydrocephalus (10%). Most reports indicate a poor prognosis with severe progressing or relapsing course when seizures are associated with sarcoidosis.[4]

In a review of the literature, among 277 patients with neurosarcoidosis, 16% had epileptic seizures, among which 50% expressed seizures as first manifestation[4].

In another study of 79 patients with neurosarcoidosis, 15% had seizures, of which 10% as first manifestation and poor prognosis[5].

Our patient falls into the category of rare EPC cases due to an inflammatory disorder and combines highly relevant features. The imaging features are in full correlation with encephalographic and clinical features of EPC[6].

The electroclinical progress set the diagnosis of subacute encephalopathy due to a possible systemic inflammatory disease in the context of neurosarcoidosis.

Based on the unfavourable development of our patient, we can conclude that when EPC appears as first manifestation of possible isolated neurosarcoidosis, it indicates poor prognosis.

According to our opinion, this case is of particular interest as an extremely rare presentation of isolated possible neurosarcoidosis with epilepsia partialis continua indicative of poor prognosis.

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