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Case Report

New onset refractory status epilepticus due to primary angiitis of the central nervous system☆

Rawan K. Matar [a,*,1], Bader Alshamsan [b], Saleh Alsaleh [c], Hindi Alhindi [d], Khalid O. Alahmedi [e], Sami Khairy [f], Salah Baz [a]

[a] Department of Neurosciences, King Faisal Specialist Hospital & Research Center, Riyadh, Saudi Arabia
[b] Department of Medicine, Qassim University, Saudi Arabia
[c] Department of Internal Medicine, King Faisal Specialist Hospital & Research Center, Riyadh, Saudi Arabia
[d] Department of Pathology, King Faisal Specialist Hospital & Research Center, Riyadh, Saudi Arabia
[e] Department of Radiology, King Faisal Specialist Hospital & Research Center, Riyadh, Saudi Arabia
[f] Department of Neurosurgery, King Abdulaziz Medical City, Riyadh, Saudi Arabia

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ABSTRACT

Primary Angiitis of the central nervous system is a rare and poorly understood variant of vasculitis. We narrate a case of a 46-year-old male who presented with new onset refractory status epilepticus mimicking autoimmune encephalitis. In this case we are reporting clues that could be useful for diagnosis and extensive literature review on the topic.

1. Introduction

New onset refractory status epilepticus (NORSE) is a complex disorder, characterized by status epilepticus that is refractory to treatment with no identifiable infectious, inflammatory or brain structural abnormalities [1]. NORSE poses considerable distress to physicians due to its heterogeneous etiology and devastating outcome [2]. An identifiable cause in patients with NORSE is discovered in the majority of cases, however in some the cause remains unknown despite extensive investigations [3]. Primary Angiitis of the central nervous system (PACNS) presenting with seizures was reported in the range of 7–29% [4,5].

PACNS is described as an entity under the "umbrella" of central nervous system (CNS) vasculitis that is confined to the central nervous system [6]. It is an extremely rare disease with an annual incidence of 2.4 cases per 1,000,000 [7]. PACNS has been reported with the greatest frequency in North America [5,8], Europe [9,10,11] and Australia [12]. Literature is lacking publications about PACNS in Saudi Arabia. The only case in the literature is of a patient with CNS vasculitis complicating a primary immunodeficiency disorder [13].

PACNS is a disease of substantial diagnostic and therapeutic challenges to clinicians [14]. Despite the efforts made to assist clinicians in the diagnosis, the decision of biopsy in a patient suspected to have PACNS remains a challenge. Here we describe the diagnostic approach, clinical characteristics, brain imaging, pathological findings and therapeutic challenges of a patient with PACNS presenting with NORSE in King Faisal Specialist Hospital and Research Center (KFSH&RC) in Riyadh. We suggest clues that could be useful for clinicians to suspect PACNS and proceed with biopsy for definitive diagnosis.

2. Case report

A 46-year-old Saudi male right-handed teacher was in his usual state of health until four months prior to his presentation. This was when he started to have a wide-based gait, unsteadiness as well as several attacks of staring and unresponsiveness. It was associated with lip smacking lasting a few seconds as noticed by his colleagues and students at school. The patient would occasionally complain of a holocranial headache lasting hours to days, incapacitating him from doing his duties of teaching and home commitments.

One week prior to his presentation to our institute, he started complaining of severe headache while in class. His colleagues directed

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* Corresponding author at: Neurology Resident, Adult Neurology, Department of Neuroscience, King Faisal Specialist Hospital and Research Center, PO BOX 3354, Riyadh 11211, Saudi Arabia.
E-mail address: rawannn@gmail.com (R.K. Matar).
[1] Permanent Address: House1587 Road926 Block1209, Kingdom of Bahrain.

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him to a local hospital. On his way he developed abnormal movements with loss of consciousness in the car. The semiology of the abnormal movements reflected left-sided tonic-clonic seizures with uprolling of the eyes, urinary incontinence, frothing of saliva that lasted few minutes and was aborted spontaneously. This was followed by a state of confusion for hours.

During the admission at the local hospital, his brother witnessed some tonic posturing of the left arm for an estimated duration of 2 minutes during which the patient was unresponsive to verbal stimuli. The patient was discharged from the local hospital without antiseizure therapy.

The patient was referred from the local hospital to our center for seizure evaluation. He presented to the Epilepsy clinic at KFSH&RC with altered consciousness and found to have aphasia. He was non-fluent, and following commands by gesture only. He was rushed to the neurophysiology clinic for an urgent electroencephalogram (EEG).

A 30-minute EEG recording revealed non-convulsive status epilepticus (NCSE). It showed a severely abnormal EEG consisting of a slow background of 6–7 Hertz (Hz) with 1–3 Hz focal slowing in the left anterior and mid temporal region. Several seizures during the recording were associated with evolution of sharp waves and spikes recurring at 3–5 Hz over the left mid and posterior temporal region. During this time the patient was unresponsive with eye blinking. This expedited his admission for investigation and management of his seizures.

He was admitted with a diagnosis of NCSE, started on intravenous (IV) phenytoin and long-term EEG monitoring. He continued to have frequent attacks of non-convulsive seizures, despite being on multiple anti-seizure drugs including lamotrigine, carbamezapine, valporic acid and pyridoxine. Long term EEG monitoring captured 13 attacks of seizures during the first 48 h. Electrographic seizure onset occurred with the onset of rhythmic sharply-contoured theta evolving to delta activity maximal in the left temporal region during unresponsiveness for 5-7 minutes.

Computed tomography (CT) brain was unremarkable for any abnormality. Lumbar puncture was performed and cerebrospinal fluid (CSF) analysis showed an elevated protein of 747 mg/dl, and white blood cells (WBC) of 51 with 94% lymphocytes. Because the CSF analysis revealed lymphocytic pleocytosis in the presence of NCSE, the patient was started on antiviral and antibiotics for possible meningo-encephalitis. CSF viral and bacterial etiologies as well as, mycobacterium tuberculosis, syphilis, brucella and sarcoidosis were ruled out.

Table 1
Serum Antibody evaluation.

A: Paraneoplastic autoantibody evaluation	
Anti-neuronal nuclear Ab, type 1	
ANNA-1, S	Negative
Reflex added	None
Anti-neuronal nuclear Ab, type 2	
ANNA-2, S	Negative
Anti-neuronal nuclear Ab, type 3	
ANNA-3, S	Negative
Anti-glial nuclear Ab, type 1	
AGNA-1, S	Negative
Purkinje cell cytoplasmic Ab type 1	
PCA-1, S	Negative
Purkinje Cell Cytoplasmic Ab Type 2	
PCA-2, S	Negative
Purkinje cell cytoplasmic Ab type Tr	
PCA-Tr, S	Negative
Amphiphysin Ab, S	Negative
CRMP-5-IgG, S	Negative
Striational (striated muscle) Ab, S	Negative
P/Q-type calcium channel Ab, S	0.00 nmol/l
N-type calcium channel Ab, S	0.00 nmol/l
ACh receptor (muscle) binding Ab	0.00 nmol/l
AchR ganglionic neuronal Ab, S	0.00 nmol/l
Neuronal (V-G) K+ channel Ab, S	0.01 nmol/l
B: Epilepsy-autoimmune antibody evaluation	
NMDA-R AB, CBA,CSF	Negative
Neuronal (V-G) K+ channel Ab, S	0.01 nmol/l
GAD65 Ab assay, S	0.00 nmol/l
GABA-B-R Ab CBA, S	Negative
AMPA-R Ab CBA, S	Negative
Anti-neuronal nuclear Ab, type 1	
ANNA-1, S	Negative
Reflex added	None
Anti-neuronal nuclear Ab, type 2	
ANNA-2, S	Negative
Anti-neuronal nuclear Ab, type 3	
ANNA-3, S	Negative
Anti-glial nuclear Ab, type 1	
AGNA-1, S	Negative
Purkinje cell cytoplasmic Ab type 2	
PCA-2, S	Negative
Purkinje cell cytoplasmic Ab type Tr	
PCA-Tr, S	Negative
Amphiphysin Ab, S	Negative
N-type calcium channel, Ab	0.00 nmol/l
P/Q-type calcium channel Ab	0.00 nmol/l
Ach receptor (muscle) binding Ab	0.00 nmol/l
AChR ganglionic neuronal Ab, S	0.00 nmol/l
CRMP-5-IgG, S	Negative

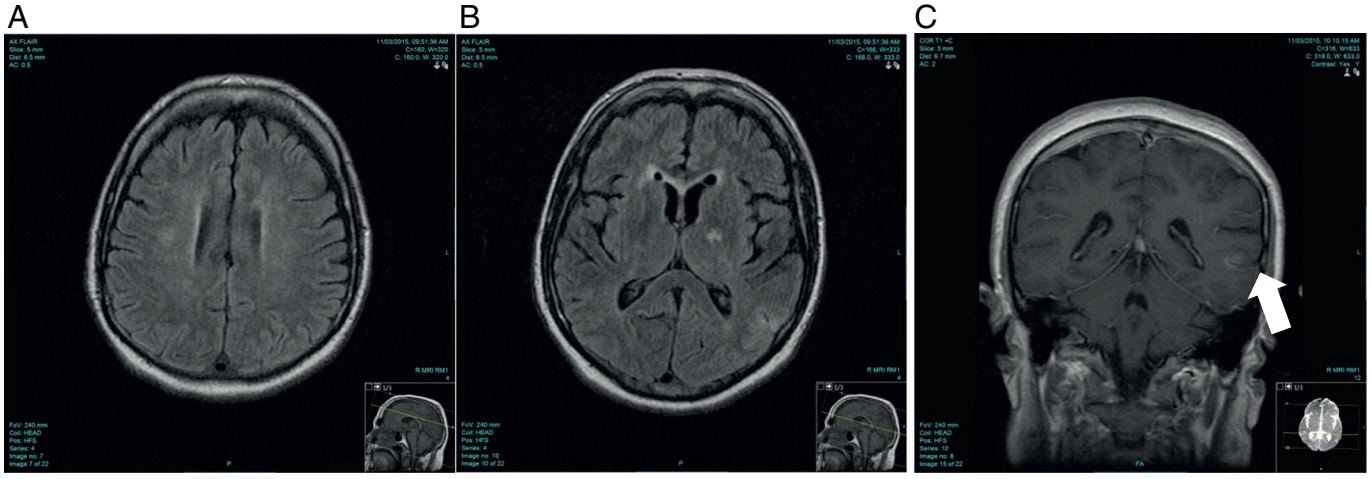

Fig. 1. MRI images at presentation A: Brain MRI FLAIR axial image showing a focal area of hyper intensity in the right peri ventricular white matter and corona radiata B: Brain MRI FLAIR axial image showing cortical and subcortical hyper intensity in the left parietal lobe C: Brain MRI T1 post contrast coronal image showing corresponding cortical enhancement of the left parietal lesion.