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Case Report

Positive Effect of Steroids in Posterior Reversible Encephalopathy Syndrome

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Posterior reversible encephalopathy syndrome · Encephalopathy · Encephalitis

Abstract
We present a case of posterior reversible encephalopathy syndrome with severe clinical manifestation. Apart from initial aphasia, hemiparesis, and a generalized seizure, the patient had a prolonged loss of consciousness. Although blood pressure was normalized, the clinical status deteriorated continuously. After adding steroids to the therapy, the patient recovered rapidly, suggesting that this could have been a useful therapeutic approach. Even the vasogenic edema in the cerebral magnetic resonance imaging disappeared shortly within 6 days.

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Introduction

Posterior reversible encephalopathy syndrome (PRES) is a severe, life-threatening neurological disorder clinically presenting with various symptoms such as headache, visual disturbances, and epileptic seizures [1]. PRES is characterized by usually bilateral cerebral vasogenic edema in computed tomography (CT) and magnetic resonance imaging (MRI) scans and occurs most commonly in patients with uncontrolled hypertensive blood pressure [2–4]. Impairment of cerebral blood flow autoregulation with consecutive local hyperperfusion is supposed to be the underlying pathomechanism [5]. Different predisposing factors such as endothelial dysfunction in the context of autoimmune diseases, sepsis, preeclampsia/eclampsia, renal failure, and toxic agents like chemotherapeutic or immunosuppressive drugs are described in the literature [6–9]. Therapeutic measurements include rigorous lowering of arterial blood pressure and treatment of complications [10, 11]. In this context we describe a patient who presented to our department with severe PRES treated with high doses of corticosteroids in addition to the arterial blood pressure lowering, resulting in rapid clinical recovery and finally complete resolution of the brain lesions in MRI.

Case Report

A 77-year-old patient with acute symptoms of left-sided middle cerebral artery stroke was referred to our department. Neurological examination revealed Broca's aphasia and right-sided hemiparesis. The initial blood pressure was 200/110 mm Hg. Cranial CT with CT angiography ruled out acute cerebral infarction as well as a cerebral hemorrhage; no arterial vessel occlusions were identified. According to current recommendations, intravenous thrombolysis with recombinant tissue-type plasminogen activator was administered. Lowering of blood pressure was immediately and successfully initiated with urapidil under constant monitoring. Nevertheless, the patient suffered a generalized epileptic seizure later on, which is why an anticonvulsive medication (with levetiracetam intravenously) was initiated. Subsequently the patient underwent cerebral MRI, revealing symmetric bilateral white matter lesions with cortical involvement in the parieto-occipital lobe and the cerebellum (Fig. 1). Diffusion-weighted MRI sequences remained unremarkable, without findings indicating acute brain ischemia in the scope of brain infarction.

Even though blood pressure was normalized in a timely manner and thrombolysis was applied, the patient's clinical status deteriorated continuously, resulting in severely impaired consciousness. In summary of the findings (clinical presentation, MRI, mild pleocytosis in cerebrospinal fluid), two differential diagnoses were considered: PRES or encephalitis. Hence, the current medication was complemented by an antiviral medication (acyclovir intravenously) and a 3-day corticosteroid treatment (1,000 mg methylprednisolone per day). Viral encephalitis caused by herpes or varicella zoster virus could not be proven, so as a logical consequence the antiviral therapy was stopped immediately. The antiepileptic medication as well as the antihypertensive therapy were continued. By that time, blood pressure remained stable in a normal range. Status epilepticus was ruled out by serial electroencephalographies. After initiation of the steroid therapy the patient's clinical status improved rapidly. After 2 days of therapy the patient completely regained consciousness. Only a mild paresis of the right arm could be noted as a residuum. After 6 days follow-up cerebral MRI demonstrated no pathological findings (Fig. 1), and no neurological deficit could be identified.

We made the final diagnosis of PRES. Retrospectively, we interpreted the brain lesions detected in the cerebral MRI as vasogenic edema as a cause of acute PRES. These lesions resolved in line with a notable clinical improvement, potentially augmented by the intravenous therapy with steroids, which were administrated due to considerations of differential diagnoses. The patient's medical history revealed rheumatoid arthritis treated with methotrexate as a preconditioning factor for PRES.

Discussion

We report the case of a patient with PRES presenting with severe clinical manifestation: aphasia, hemiparesis, generalized seizure, and prolonged loss of consciousness. Although blood pressure was normalized, the clinical status deteriorated continuously. After adding steroids to the therapy, the patient recovered rapidly, suggesting that this could have been a useful therapeutic measure. Even the vasogenic edema in the cerebral MRI disappeared within few days.

PRES usually presents with symptoms such as headaches, seizures, and focal neurological deficits that recede on average 8 days after the onset of symptoms; in 10–20% of patients residual neurological deficits persist [5, 12, 13]. Our patient experienced obviously a rare form of PRES with severe clinical presentation; in 1 out of 10 patients a persisting loss of consciousness leads to the necessity for oral intubation and artificial ventilation [13]. Severe forms of PRES are associated with a higher mortality and are less reversible [12, 14, 15]. After 3 months half of the patients with severe PRES still have a functional impairment [2]. In our case, further complications could be avoided as the clinical status improved, underlining the hypothesis of a positive effect of high-dose steroids in the acute phase of the disease.

Considering the efficiently normalized blood pressure in our patient in presence of his severe clinical condition before starting steroids, the effect on the positive outcome due to the latter seems likely. The short-term recovery in our patient within less than 2 days – from prolonged loss of consciousness to only mild paresis of the right arm – also suggests that steroid therapy potentially boosted the recovery process.

In line with the clinical improvement the structural changes depicted in the brain imaging disappeared after 1 week. In the follow-up examination no pathological findings suggesting a vasogenic edema could be identified. Reviewing the literature, 1 month after the index event, more than half of the patients present with pathological findings related to PRES in the cerebral MRI [12]. In severe cases of PRES residual lesions might occur even more often [16]. In this context the complete resolution of brain lesions after 1 week in our patient also supports the hypothesis that the administration of steroids significantly contributed to this development.

Considering the potential mechanism in PRES, with disturbance of cerebral blood flow and consecutive development of a vasogenic edema, it seems logical that therapeutic measures are aimed at the treatment of this edema. It is well known that steroids have a favorable influence regarding the outcome in different brain disorders associated with secondary edema [17–21]. Therefore and based on pathophysiological considerations, a positive treatment effect of steroids for reducing edema caused by PRES appears likely. Our case report might support this hypothesis. However, systematic investigations are needed to verify the effect of steroids in acute PRES.

Statement of Ethics

The patient gave informed consent for publication of this case report.

Disclosure Statement

The authors declare that they have no competing interests.

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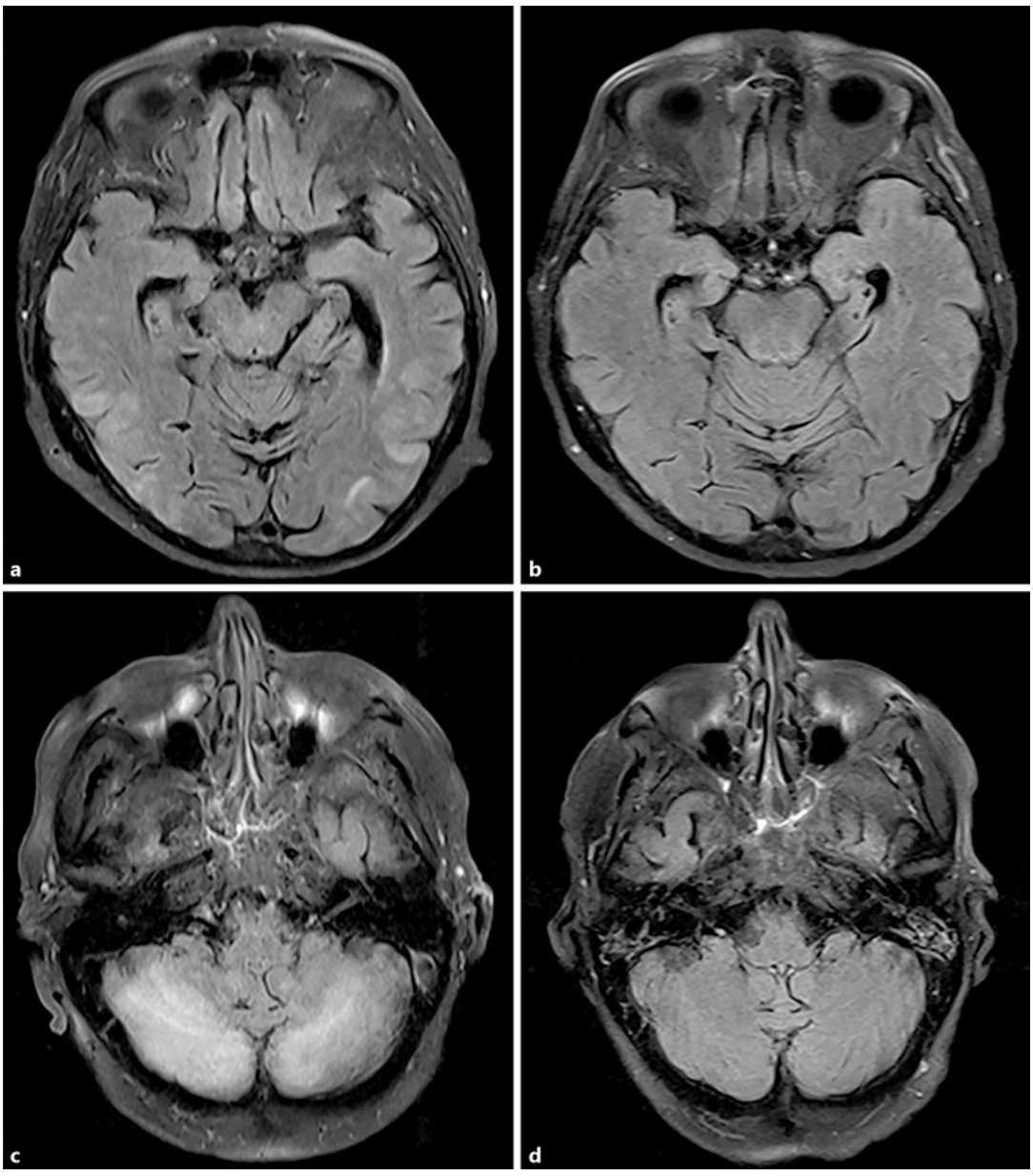

Fig. 1. **a**, **c** Vasogenic edema in the parieto-occipital lobe and cerebellum in axial FLAIR magnetic resonance images. **b**, **d** Completely regressive vasogenic edema in axial FLAIR magnetic resonance imaging after consequent blood pressure lowering and corticosteroid treatment.