CASE REPORT

PRES with asymptomatic spinal cord involvement. Is this scenario more common than we know?

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INTRODUCTION: Posterior reversible encephalopathy syndrome (PRES) is an entity characterized by neurologic symptoms such as headaches, altered mental status, seizures and visual changes, and it is associated with white matter vasogenic edema predominantly affecting the posterior occipital and parietal lobes of the brain.

CASE REPORT: A 19-year-old patient developed PRES after the use of chemotherapy for a testicular teratocarcinoma and after the development of a blood pressure elevation.

DISCUSSION: Few cases described the involvement of the spinal cord in this syndrome. In the majority of these cases, the spinal cord involvement was asymptomatic or with few symptoms of spinal cord disease.

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INTRODUCTION

Posterior reversible encephalopathy syndrome (PRES) is a clinicroadiologic syndrome that is characterized by headaches, altered mental status, seizures and visual disturbance, and it is associated with white matter vasogenic edema predominantly affecting the posterior occipital and parietal lobes of the brain.¹

Many factors can act as triggers of this syndrome; the most common factors are the acute elevation of blood pressure, such as during eclampsia, abnormal renal function and immunosuppressive therapy.^{1,2} Other possible etiologies already described are autoimmune diseases (lupus)³, allogeneic bone marrow and solid organ transplantation^{4,5}, neoplasia and chemotherapy treatment^{6,7}, systemic infections, sepsis^{8,9} and exposure to venom.^{3–10}

The pathophysiology of PRES remains unclear; however, it appears to be related to cerebral autoregulation failure. The cerebral autoregulation maintains the cerebral blood flow constant over a range of mean blood pressures by arteriolar vasoconstriction and vasodilatation.¹¹ If the upper limit of cerebral autoregulation is exceeded, the arteriolar system will dilate leading to brain hyperperfusion and blood–brain barrier disruption, allowing a cerebral edema.^{11,12}

The most common radiological presentation of PRES is an increase of signal in T2-fluid-attenuated inversion recovery (FLAIR) in both parieto-occipital lobes.^{1,2} There are a few cases in literature that describe the spinal cord involvement in this syndrome.

CASE REPORTS

We report the case of a 19-year-old patient with a history of testicular teratocarcinoma treated with local surgery and BEP (bleomycin, cisplatin and etoposide) chemotherapy who was submitted to an extensive resection of paraaortic lymph nodes. After this procedure, he developed a stenosis in his right renal artery and secondary hypertension. On admission, his creatinine level raised to 2.35 mg dl⁻¹, and it came back to normal values a week later. Two months later he came to the emergency room

complaining of headache, nausea and a central scotoma in the left eye. On examination, blood pressure levels were 180/140 mm Hg, heart rate was 85 and his temperature was normal. Partial third-nerve palsy was evident, along with gait ataxia and bilateral optic disc edema, retinal hemorrhages and vasospasm. Mental status and muscular strength were unremarkable.

Laboratory studies showed a creatinine value of 1.01 mg dl⁻¹, and undetectable B-HCG and alpha-fetoprotein. His laboratory test results were as follows: uric acid, 4.6 mg dl⁻¹; LDH, 614 U l⁻¹; sodium, 138 meq l⁻¹; and potassium, 3.6 meq l⁻¹. Initial brain CT scan did not show any abnormalities.

He was admitted and was started on intravenous antihypertensive drugs to control the severely elevated blood pressure.

We performed brain magnetic resonance imaging (MRI) studies that demonstrated bilateral basal ganglia and brain stem increase of signal in FLAIR, suggestive of PRES. The brain MRI also showed a cyst in the left temporal lobe (Figure 1). Because of involvement of the brain stem, we performed a cervical spinal cord MRI to evaluate the extension of the lesion, and this MRI showed an increase of signal in T2 in the interior of the cervical spinal cord. (Figure 2) Despite extensive edematous lesions on the spinal cord, he remained without sensitive and motor deficits. After his BP and creatinine peaked to 240/140 and 1.54 mg dl⁻¹, respectively, his condition started to improve quickly, which completely resolved his neurological symptoms.

Two weeks later a control MRI did not show any permanent lesion. Nowadays, 6 months after the episode, he is currently on blood pressure medications and remains asymptomatic.

DISCUSSION

After Hinchey's paper in 1996, many new publications had reported clinical situations and drugs that can be directly or indirectly associated with PRES.¹ Nowadays, PRES is a clinicroadiologic entity encountered in several clinical conditions. The increase in PRES descriptions in literature is becoming more common because of the elevation of the number of patients in

Figure 1. Brain MRI showing an increase of signal in T2/Flair in bilateral basal ganglia and brain stem.

Figure 2. Cervical MRI demonstrated an increase of signal in T2 in the interior of the spinal cord.

chemotherapy and in the use of immunosuppressive agents, as well as the widespread of MRI technology.

The most characteristic imaging pattern in PRES is the presence of an edema involving the white matter of the posterior portions of both cerebral hemispheres, especially the parieto-occipital regions, in a relatively symmetric pattern that spares the calcarine and paramedian parts of the occipital lobes. However, other structures such as the brain stem, cerebellum and frontal and temporal lobes may also be involved, and although the abnormality primarily affects the subcortical white matter the cortex and the basal ganglia may also be affected. The reversibility of the clinical manifestation and the improvement in the neuroimage exams are 'hallmarks' of the PRES.^{1,8,13} There are a few cases in literature that report the involvement of the spinal cord in this syndrome.^{14–18}

The elevation of blood pressure was reported in the cases of spinal cord involvement in PRES. In the case described in this paper, the patient was exposed to many triggers for PRES: elevation of blood pressure, acute renal failure and chemotherapy.^{14–18}

Another interesting topic is that PRES involves predominantly the white matter of the brain, and in the brain stem and spinal cord the lesions described affect the gray matter. Normally, patients with spinal cord lesion had alterations in neurological examination (sensitive and motor deficits). However, in this case and in other previously described cases, there were no neurological deficits related to spinal cord lesion. There is only one case that describes a neurological signal (Babinski sign) in a patient with spinal cord involvement.^{14–17}

The prediction for posterior brain regions is not yet understood. Probably, the low sympathetic innervation in posterior circulation leads to the prediction for this topography.¹⁹ Typically, PRES occurs predominantly in vertebro-basilar artery topography

distribution. The cervical spinal cord is irrigated by the anterior spinal arteries (that are branches of vertebral arteries).²⁰

In the experience of our group, we evaluated 51 patients during the past 5 years and this is the first case of spinal cord injury in patients with PRES.

In the majority of these cases, the spinal cord involvement was asymptomatic or with few symptoms of spinal cord disease. We speculate that the involvement of the spinal cord is more common than we see described in literature, because spinal cord involvement leads to none or few symptoms, and there are few patients with PRES who were submitted to cervical spinal cord MRI.

COMPETING INTERESTS

The authors declare no conflict of interest.

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