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Review

Hypertensive posterior reversible encephalopathy syndrome causing posterior fossa edema and hydrocephalus

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ABSTRACT

Posterior reversible encephalopathy syndrome (PRES) is a well characterized entity resulting from the inability of cerebral autoregulation to adequately protect the brain from uncontrolled hypertension. It primarily affects the occipital lobes, but can also involve the structures in the posterior fossa including the brainstem and cerebellum. Treatment usually consists of strict blood pressure control, but more aggressive management may be indicated with acutely worsening neurological status. We present a patient with hypertensive encephalopathy that resulted in hydrocephalus and brainstem compression necessitating surgical decompression requiring ventriculostomy and suboccipital craniectomy. In rare cases, PRES can present with severe brainstem compression requiring emergent posterior fossa decompression. When brainstem signs are present on exam, emergent posterior fossa decompression may be safer than ventriculostomy alone.

1. Introduction

Hypertensive encephalopathy, also known as reversible posterior leukoencephalopathy syndrome or posterior reversible encephalopathy syndrome (PRES), is characterized by headaches, lethargy, altered mental status, visual complaints, or seizures [1,2]. These symptoms result from blood pressure exceeding the upper limit of cerebral autoregulation, which leads to breakdown of the blood–brain barrier and extravasation of fluid and protein into the brain parenchyma [2–4]. Hypertensive encephalopathy primarily affects the subcortical white matter of the occipital lobes [2]. Hypertension leading to edema involving the cerebellum is less common, but there are several reports in the literature [2,4–6]. Most cases of hypertensive encephalopathy resolve with blood pressure management [4–6], with some requiring external ventricular drainage [2]. Unsuccessful weaning of the ventriculostomy may require ventricular shunting [7]. We report a patient with hypertension-induced cerebellar edema leading to hydrocephalus, compression of the brainstem, and cranial nerve deficits requiring emergent surgical decompression.

2. Case report

2.1. History and physical examination

A 65-year-old woman with history of colon cancer was found unresponsive in her home in December 2011. She was intubated by emergency medical services and brought to an outside hospital. A head CT scan demonstrated cerebellar edema with compression of the fourth ventricle, effacement of the quadrigeminal, perimesencephalic, and prepontine cisterns, compression of the brainstem, and associated hydrocephalus involving the lateral and third ventricles with transependymal flow (Fig. 1). She was transferred emergently to the University of Iowa Hospital for further management.

On arrival, the patient's blood pressure was 217/113 mmHg. She had no history of hypertension. She was intubated without sedation or paralytics. Her right pupil was 3 mm and unreactive. The left pupil was 4 mm and minimally reactive. She had weak corneal reflexes bilaterally. She was not initiating breaths on the ventilator, nor did she demonstrate cough or gag reflexes. She did not move her upper extremities to central noxious stimulus. She was hyperreflexic throughout, had a triple flexion response in her lower extremities bilaterally, and had positive Hoffmann's reflex bilaterally.

2.2. Operation

Given her poor neurological examination and evidence of brain stem compression, the patient was taken emergently to the operating room for suboccipital craniectomy and external ventricular drain placement. Initially, diagnoses of cerebellar neoplasm or infarction were entertained. In the operative room a standard right frontal ventriculostomy was placed. A midline suboccipital craniectomy was then performed spanning from the transverse sinus rostrally to the foramen magnum caudally. Upon durotomy, the cerebellar tissue was noted to be under high pressure.

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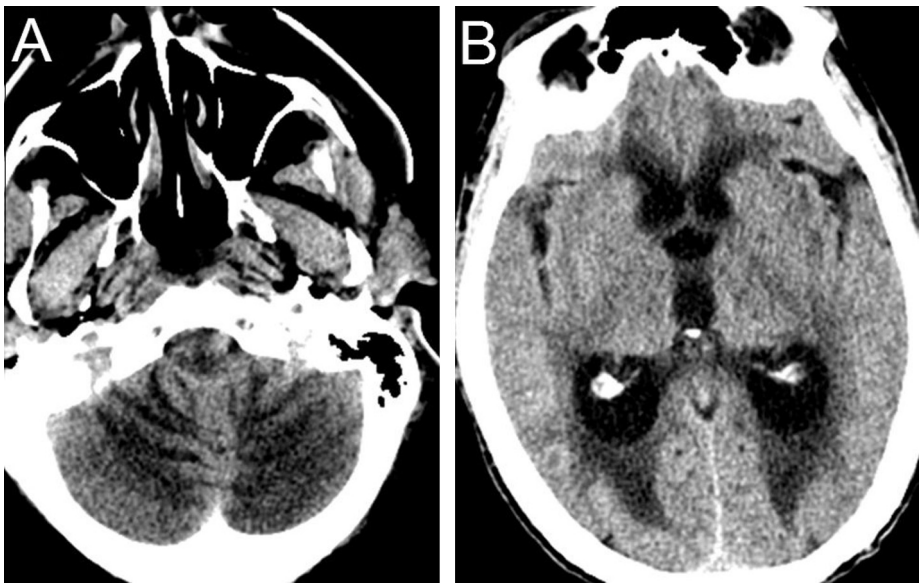

Fig. 1. Pre-operative non-contrast axial head CT scan showing (A) cerebellar edema, effacement of fourth ventricle and basilar cisterns, and compression of the brainstem, and (B) dilated lateral ventricles with transependymal flow.

Intra-operative ultrasound was performed, but there was no evidence of mass lesion. Biopsy was not performed. The patient tolerated the procedure well and was admitted to intensive care post-operatively.

2.3. Post-operative course

On post-operative examination, the patient's pupils were 4 mm, equal, and reactive bilaterally. She flexed her upper extremities bilaterally to central noxious stimuli. She also demonstrated eye opening to noxious stimuli. A combination of intravenous hydralazine and labetalol was used to maintain systolic blood pressure below 160 mmHg. Mannitol 20 g every 6 hours was initiated, and weaned over several days. Cerebrospinal fluid analysis from the time of surgery was not concerning for infectious process. Additional laboratory work was largely unrevealing, but it did show that the patient had hypothyroidism and she was started on thyroid replacement therapy. Oncologic work-up with CT positron emission tomography (PET) was negative confirming good systemic control of her colon cancer. Echocardiogram was significant only for mild left ventricular hypertrophy.

MRI obtained immediately post-operatively demonstrated diffuse cerebellar edema along with T2-weighted hyperintensities of the periventricular and occipital white matter (Fig. 2). There was no mass lesion or restricted diffusion suggestive of acute infarction on MRI. Head CT scan on post-operative day 4 demonstrated good decompression of the posterior fossa with improvement of the hydrocephalus along with partial resolution of the cerebellar edema (Fig. 3). There was a small amount of hemorrhage along the ventriculostomy tract with layering of blood in the occipital horns of the lateral ventricles.

Ventriculostomy was weaned over the course of 7 days. The patient continued to improve neurologically throughout her hospitalization. Prior to discharge she was awake and alert with mild confusion. Cranial nerves were intact. She followed commands with 5/5 motor strength in all extremities and had intact sensation. Reflexes were normal with downgoing toes bilaterally.

At her 6 month follow-up the patient reported a return to baseline functioning. Her modified Rankin scale score was 0. She complained of occasional mild headaches that were much improved since discharge. Her blood pressure had been well controlled on oral antihypertensives. Neurological exam showed the patient was awake, alert, and oriented to time, place, and person. Cranial nerves were intact. Reflexes were normoreflexic throughout and there was no Hoffmann's reflex, no clonus, and no Babinski reflex. Gait and coordination were normal. MRI obtained at follow-up showed resolution of the cerebellar edema with significant improvement of the hydrocephalus (Fig. 4). There were some residual periventricular white matter T2-weighted hyperintensities.

3. Discussion

3.1. Hypertensive encephalopathy

Blood flow to the brain is maintained over a relatively wide range of blood pressures by cerebral autoregulation via dilation and constriction of cerebral arterioles [1–4,8]. Chronic hypertension may increase the upper limit of autoregulation [3]. If the blood pressure becomes too high, it exceeds the brain's ability to autoregulate blood flow. This leads to increased arteriolar and capillary pressures resulting in breakdown of the blood–brain barrier and extravasation of fluid and protein into the brain parenchyma [2,4]. It has been proposed that the greater density of sympathetic innervation of the anterior cerebral circulation relative to the posterior circulation leads to the preservation of autoregulation and therefore the blood–brain barrier in the former, while that of the latter fails [2,3,9]. This may explain why hypertensive encephalopathy preferentially affects the occipital lobes [1–3].

Most patients with hypertensive encephalopathy, including cases involving the cerebellum and brainstem, can be managed simply with control of the blood pressure [4–6,9]. Patients with severe symptoms secondary to hydrocephalus from obliteration of the fourth ventricle may need ventricular drainage [2,7]. In such patients, ventriculostomy can usually be weaned successfully, but permanent drainage may be necessary [7]. Osmotic diuretics such as mannitol may be helpful in the face of cerebral edema, however clinical evidence regarding the use of mannitol when dealing with hypertensive encephalitis is lacking. The use of steroids such as dexamethasone has been reported by some authors [2,5], but evidence of its efficacy is similarly lacking. There are also