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Case Report

Hypertensive posterior reversible encephalopathy causing obstructive hydrocephalus

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Received : 19 October 2022
Accepted : 03 March 2023
Published : 17 March 2023

DOI
10.25259/SNI_963_2022

Quick Response Code:

ABSTRACT

Background: Posterior reversible encephalopathy syndrome (PRES) can occur due to the detrimental effect of malignant hypertension on cerebral autoregulation. Most reported cases describe involvement of the supratentorial areas. Involvement of the posterior fossa structures in conjunction with supratentorial involvement has also been reported; however, PRES affecting the infratentorial structures without supratentorial involvement is a rare phenomenon. Clinical manifestations can involve severe headache, seizures, and reduced consciousness with treatment focused primarily on blood pressure control.

Case Description: We report a case of PRES with isolated involvement of the infratentorial structures leading to obstructive hydrocephalus. The patient was managed with aggressive control of blood pressure and avoided ventriculostomy or posterior fossa decompression with a good outcome.

Conclusion: Medical management in the absence of neurological deficit can be associated with a good outcome.

Keywords: Cerebral autoregulation, Hypertensive encephalopathy, Obstructive hydrocephalus, Posterior fossa oedema

INTRODUCTION

Posterior reversible encephalopathy syndrome (PRES) or hypertensive encephalopathy is an entity associated with hypertensive crisis and typically presents with acute headache, seizures, or disorders of consciousness.[4] Other conditions that are implicated in the development of PRES include (pre) eclampsia, cytotoxic mediation, and renal disease but discussion of these are beyond the scope of this report.[4] Posterior fossa PRES has been reported in the literature, generally in association with supratentorial involvement. However, isolated infratentorial PRES causing hydrocephalus is a rare phenomenon.[5] Cerebellar edema from several etiologies including infarction or tumor can compress the fourth ventricle or cerebral aqueduct and cause obstructive hydrocephalus. Although hypertensive encephalopathy predominantly affects the subcortical white matter of the occipital lobes, a similar process leading to diffuse vasogenic edema in the posterior fossa may result from significant hypertension.[3] In this setting, emergent CSF diversion with an external ventricular drain or posterior fossa decompression may be required.[5,7,11] However, in a small number of cases including ours, aggressive treatment of underlying hypertension can lead to reversibility of the condition without surgical intervention.[9]

We present a case of posterior fossa PRES due to hypertensive crisis that presented with acute obstructive hydrocephalus and discuss pertinent clinical and radiological features.

Literature search

A search of PubMed and Medline databases was reviewed for articles from inception to April 2022 inclusive. Limits were placed on those articles published in the English language. We excluded editorials, commentaries, letters, conference papers, and reviews. Our search only identified case reports; therefore, we did not perform a systematic review.

CASE REPORT

A 39-year-old male, Caucasian, smoker who was usually fit and well presented with a 4-month history of progressive headache and new onset vomiting leading to attendance at the emergency department. Headaches were suggestive of raised intracranial pressure, worse on lying flat and bending forward. The patient reported blurriness of his vision when the headaches were severe, but there were no lower cranial nerve signs. Blood pressure on arrival was 241/151 mmHg. Fundoscopy demonstrated Grade III papilledema in the right eye. The rest of the neurological examination was normal. An urgent computed tomography head scan demonstrated mild hydrocephalus and possible diffuse cerebellar edema, but no mass lesion or hemorrhage [Figure 1]. The patient was commenced on antihypertensive medications (amlodipine and bisoprolol) before a magnetic resonance imaging (MRI) scan was performed. MRI demonstrated T2 hyperintensity involving the cerebellum, midbrain, pons, and medulla [Figure 2]. Cerebellar edema was once again demonstrated. Resultant effacement of the fourth ventricle was seen with hydrocephalus and evidence of periventricular hyperintensity. There were also multiple small T2 hyperintensities in the supratentorial white matter, suggestive of microvascular changes. Microhemorrhages were evident within the left cerebellar hemisphere, brainstem, putamen, and temporal lobe; deemed to be due to profound hypertension. There was no restricted diffusion or abnormal enhancement in relation to the abnormalities described above. The patient was admitted under neurosurgery and underwent frequent neurological examinations in case of any deterioration requiring surgical intervention. As the patient remained Glasgow coma scale 15; the hydrocephalus was managed conservatively and aggressive control of blood pressure was undertaken. Investigations for secondary causes of hypertension were negative. Following optimization of blood pressure with oral calcium channel blockers, the patient was subsequently discharged as clinical symptoms had improved. Repeat MRI imaging 6 weeks post presentation (and before discharge) demonstrated resolution of the posterior fossa edema and hydrocephalus [Figure 3]. The patient remained well with complete resolution of headaches at outpatient review 6 months later.

DISCUSSION

Posterior fossa PRES presenting with acute hydrocephalus is a rare phenomenon. The pathogenesis of PRES is poorly understood but may involve cerebrovascular endothelial dysfunction resulting in impaired autoregulation, vasodilation, and disruption of the blood-brain barrier leading to vasogenic edema.[6,12] Endothelial cell injury and extravasation of protein and fluid into the parenchyma could be implicated in the pathogenesis of this condition. One hypothesis for increased tendency of PRES affecting the posterior circulation, is that greater density of sympathetic innervation of the anterior cerebral circulation allows for tolerance of a greater insult and maintenance of cerebral autoregulation compared with the posterior circulation.[12,13]

Hypertensive encephalopathy can affect both supra- and infratentorial structures. Cerebellar edema resulting from

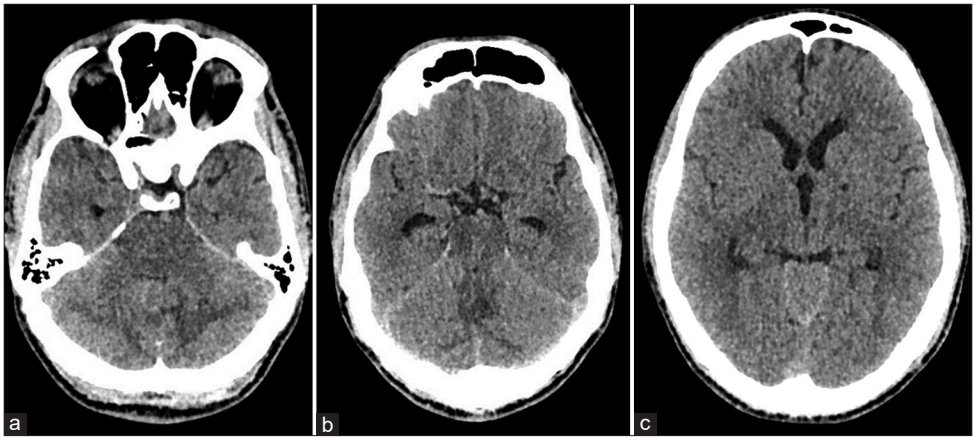

Figure 1: Initial noncontrast axial computed tomography head scan demonstrating (a) cerebellar oedema with effacement of the fourth ventricle, (b and c) corresponding supratentorial ventriculomegaly.

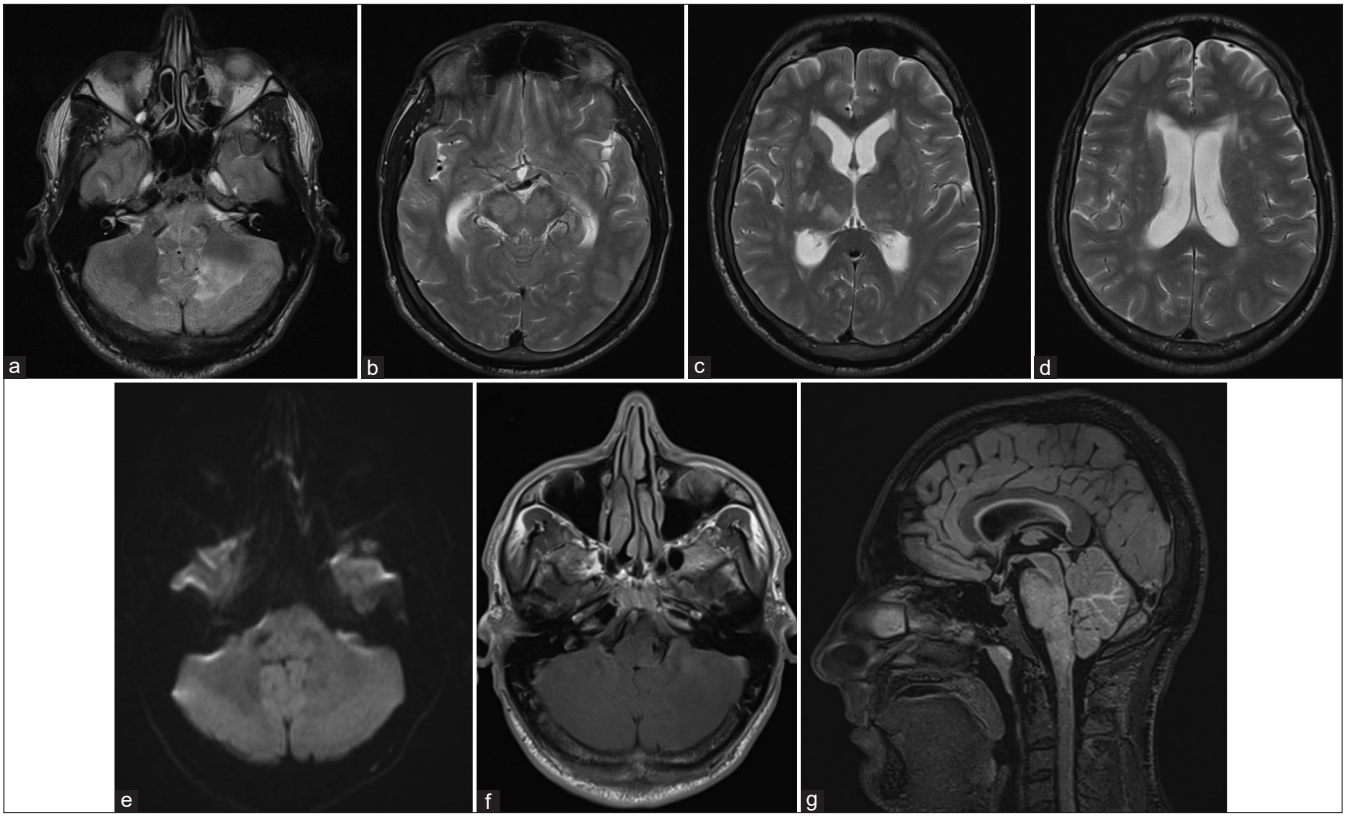

Figure 2: Initial magnetic resonance imaging showing T2/fluid attenuated inversion recovery hyperintensities in the cerebellum and brainstem (a, b, and g) and dilated lateral ventricles with periventricular hyperintensity (c and d). No infarction on diffusion-weighted imaging (e) or enhancing lesion on contrast-enhanced T1 imaging (f).

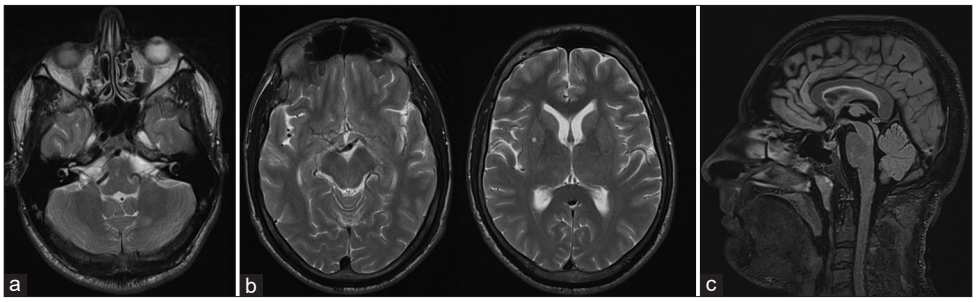

Figure 3: Magnetic resonance imaging 6 weeks later demonstrating resolution of signal change and swelling of the infratentorial structures and improvement of hydrocephalus (a and c). Occasional T2 hyperintensities in supratentorial white matter persist, presumably due to mild small vessel disease (b).

PRES is a known entity, but this is often associated with concomitant supratentorial edema.[14] In rare cases, disease can be isolated to the posterior fossa structures and resultant edema can cause fourth ventricular or aqueduct compression leading to obstructive hydrocephalus. In other reports, patients with posterior fossa PRES may present with profound depression of consciousness which necessitates ventriculostomy or posterior fossa decompression.[1,5] In our case, aggressive control of blood pressure led to reversal of hydrocephalus and improvement of headaches without the need for ventriculostomy or surgical decompression. It is unclear what factors are responsible for variation in the severity of presenting symptoms in hypertensive encephalopathy of the posterior fossa.

An important distinguishing feature of posterior fossa PRES may be the clinical-radiological dissociation that we observed, which corroborates with findings elsewhere.[2] Diffuse cerebellar and brainstem edema with relative paucity of brainstem signs in the context of significant uncontrolled hypertension may distinguish hypertensive encephalopathy from other structural lesions that can affect the posterior fossa.[10] Despite extensive radiological findings of increased

fluid attenuated inversion recovery signal/edema [Figure 2] in the brainstem, the patient did not exhibit any profound cranial nerve deficits or motor weakness. In our case, diffusion-weighted imaging did not demonstrate restricted diffusion, as has also been observed in other studies.[8]

CONCLUSION

This report highlights that hypertensive encephalopathy of the posterior fossa can present with obstructive hydrocephalus which may resolve with aggressive control of blood pressure. However, close monitoring and a threshold for rapid decompression must remain in the treatment armamentarium. Radiological findings that manifest with this condition warrant awareness as a differential, as prompt recognition and treatment of underlying significant hypertension can be associated with good outcomes. Further larger scale studies are needed to elucidate this clinical syndrome further, particularly with respect to predictive factors for the varying degree of clinical presentations.

Statement of ethics

Patient consent for publication was obtained.

Author contributions

SM: Drafting and reviewing manuscript. SA: Reviewing and editing the manuscript. NS: Reviewing the manuscript. CD: Reviewing the manuscript.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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How to cite this article: Moughal S, Ahmad S, Saleem N, Derham C. Hypertensive posterior reversible encephalopathy causing obstructive hydrocephalus. Surg Neurol Int 2023;14:94.