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Hindsight is 20/20, an unexpected ocular telltale sign of bilateral renal artery dissection

Liyung T. Chou, Azin Abazari *

Stony Brook University Medical Center, Department of Ophthalmology, HSC L2, Rm 152, Stony Brook, NY, 11794-8223, USA

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1. Case report

A 42-year-old healthy male presented to the emergency department with rapid binocular vision loss, back pain, and neck pain. Two weeks prior, he had similar backpain and was treated at an outside hospital for suspected pyelonephritis versus nephrolithiasis with antibiotics and pain medication. The patient denied any prior ocular or medical history. His blood pressure was 159/109 mmHg (unknown baseline but was reportedly normal) with an abnormal kidney function test and hematuria (Cr 1.7 (0.5–1.20, unknown baseline), UA with large number of red blood cells (RBC) and three white blood cells (WBC)). Non-contrast computed tomography (CT) of the head and CT angiogram of the neck were normal. Ophthalmology was consulted given concern for non-organic cause of vision loss. On exam, his vision was light perception in both eyes. Pupils were briskly reactive to light without afferent pupillary defect. Anterior segment and fundus exam were unremarkable without cotton wool spots, arterial narrowing, tortuosity, flame shaped hemorrhages, or cherry red spot. The optic nerves were pink without disc edema. Optokinetic testing did not show any response, indicating vision worse than 20/400. The patient had a normal neurologic exam with intact mental status without ataxic gait, nystagmus, and saccadic intrusions. Finally, brain magnetic resonance imaging (MRI) with and without contrast revealed multiple T2/FLAIR hyperintensities in the bilateral occipital and cerebellar lobes. There were additional T2/FLAIR hyperintensities in juxtacortical region of the bilateral frontal parietal lobes involving the central sulcus with associated leptomeningeal enhancement (Fig. 1). Given that only one WBC was noted on the lumbar puncture, the enhancement was less likely due to an infectious process, and the overall radiologic findings were suggestive of posterior reversible encephalopathy syndrome (PRES). As the patient was hypertensive with systolic blood pressure in the 200's, CT angiography of abdomen was done and revealed bilateral renal artery infarcts/thrombosis (Fig. 2). Renal artery duplex ultrasound subsequently demonstrated possible dissection and a surprising bilateral spontaneous renal artery dissection (SRAD) was eventually confirmed by renal arteriogram. The patient was treated with bilateral renal artery stents with normalization of his blood pressure and vision. At three-month follow-up, his visual acuity was 20/20 in both eyes and the T2 FLAIR hyperintensities on MRI resolved (Fig. 3). Workup for hypercoagulable syndrome, connective tissue disease, and vasculitis was unrevealing and included titers for lupus anticoagulant, anti-cardiolipin antibody, antithrombin III, protein C and S, factor V Leiden, ANA, ANCA, and dsDNA. ESR was 48 (0–15 mm/h) and CRP was 4.6 (0–0.3 mg/dL). Urine drug screen was negative.

2. Discussion

PRES is a clinical-radiological entity often characterized by headache, encephalopathy, seizure, and visual disturbance such as decreased vision, visual field defects, or cortical blindness despite an unremarkable eye exam. Binocular diplopia, though rare, has also been reported. PRES is associated with multiple conditions including hypertension, renal failure, preeclampsia and eclampsia, autoimmune conditions, immunosuppression, and most recently, COVID-19.[1] PRES is also known as reversible posterior leukoencephalopathy syndrome; however, both terminologies are somewhat a misnomer as the condition is not always reversible especially with delayed recognition. While the incidence of

* Corresponding author.
E-mail address: azin.abazari@tuftsmedicine.org (A. Abazari).

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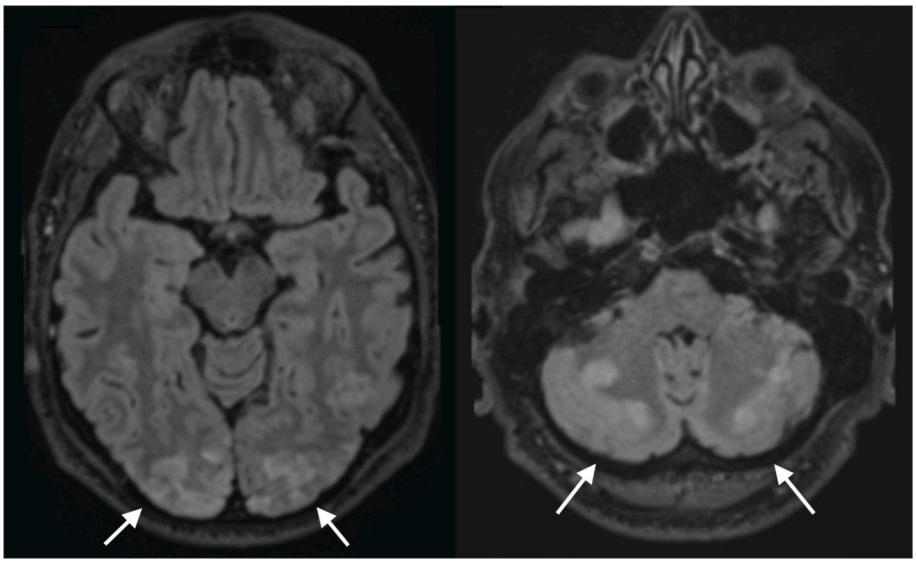

Fig. 1. Magnetic Resonance Imaging - Brain. Multiple T2/FLAIR hyperintensities in bilateral occipital lobes (left, arrows) and cerebellar lobes (right, arrows) consistent with PRES.

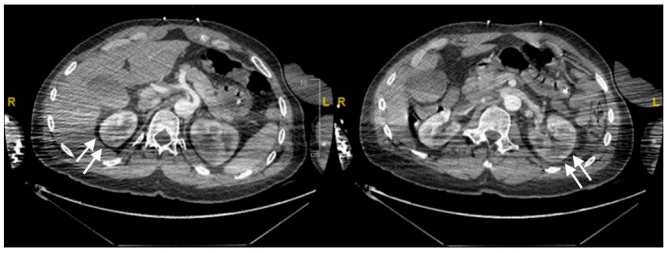

Fig. 2. Computed Tomography Angiography – Abdomen. Nonenhanced areas involving the superior pole of the right kidney parenchyma (left, arrows), and multiple areas of nonenhancement involving the middle pole of the left kidney (right, arrows) consistent with infarcts.

PRES in the general population is unknown, it has been reported to occur in 0.84% of patients with end stage renal disease, 0.69% with systemic lupus erythematosus, 2.7–25% with bone marrow transplantation, and 0.4% with solid organ transplantation. Typical radiologic features include T2 hyperintense vasogenic edema in the parietal-occipital hemisphere that usually resolve upon recovery. The pathophysiology remains unclear, but it has been postulated that the posterior hemisphere is more susceptible to the insult due to less sympathetic innervation in the posterior hemisphere protecting against autoregulation dysfunction and hyper-perfusion. Another possibility is that the affected regions are the watershed areas of the brain, making them prone to vascular changes. It is worth noting that reversible cerebral vasoconstriction syndrome (RCVS) is closely related to PRES and should be kept in the differential diagnosis. Nevertheless, RCVS can be distinguished by thunderclap headache and less commonly associated focal neurologic deficits, stroke, and seizure. Brain imaging findings are different and typically demonstrates reversible narrowing of cerebral arteries. Despite PRES being a well-known entity, it remains underdiagnosed given its

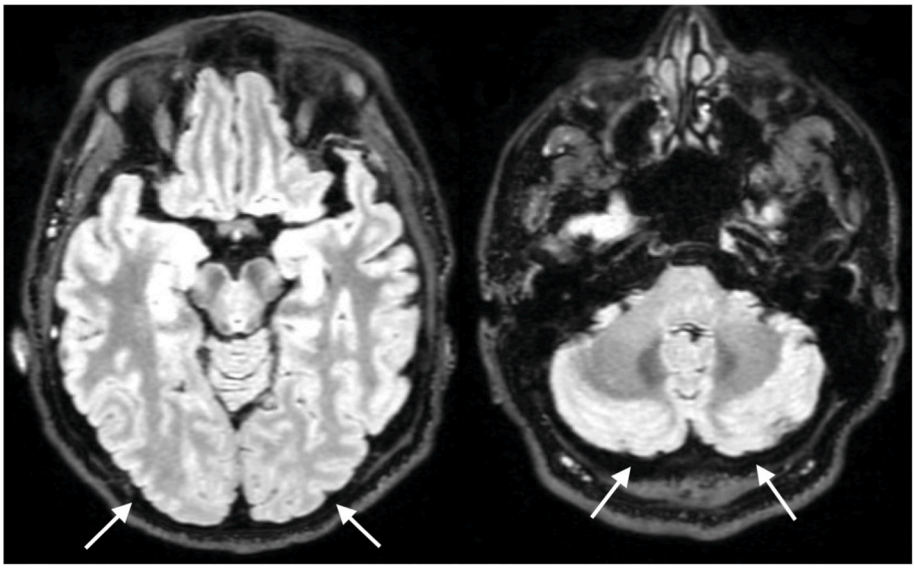

Fig. 3. Magnetic Resonance Imaging – Brain. Previously seen FLAIR hyperintensities in bilateral occipital lobes (left, arrows) and cerebellar lobes (right, arrows) have completely resolved as patient recovered from PRES.

variable clinical presentations.[2] For our patient, non-organic cause of vision loss was entertained and assessed with optokinetic testing given the initial vague combination of visual changes, hypertension, and renal abnormality (though our patient did eventually develop seizure after diagnosis of PRES). Nevertheless, since as high as 9.9% of patients have vision loss in PRES,[2] it is important for ophthalmologists to obtain detailed history, attend to vital signs, and consider pathology in the retro-orbital visual pathway when there is no evidence of ocular abnormality. Indeed, visual changes is a poor diagnostic criterion for PRES and a high clinical suspicion is needed.[2,3] Certainly, while PRES can be reversible, it is not always the case and can in fact lead to death in 3–6% of cases secondary to intracranial hemorrhage, increased intracranial pressure, and hydrocephalus or brain stem compression from cerebral edema.[1] As a result, PRES requires prompt workup and timely management of the underlying condition. For our patient, recognizing the renal artery dissection early prevented renal failure and the subsequent irreversible complications of PRES from a persistent cerebral autoregulation dysfunction.

3. Conclusion

PRES remains difficult to diagnose, despite known associations, given its variable clinical presentations. Ophthalmologists should consider PRES in patients with visual loss localized to the retrogeniculate visual pathway and accompanied neurologic findings as early diagnosis and treatment of the underlying condition is critical in ensuring favorable prognosis.

4. Patient consent

The patient(s)/patient's legal guardian consented to publication of the case in writing/orally.

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Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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