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Reversible Vasoconstriction Syndrome with Bilateral Basal Ganglia Hemorrhages

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Abstract

Reversible cerebral vasoconstriction syndrome (RCVS) is an increasingly recognized acute cerebrovascular condition that may produce myriad transient and sustained neurologic deficits as well as a host of radiologic features. We report the case of a woman with RCVS and a severe clinical syndrome with bilateral basal ganglia hemorrhages, cerebral infarctions, and marked vascular abnormalities. The patient made a near complete clinical recovery, representing an extreme and illustrative form of RCVS.

Keywords

Stroke; hemorrhage; vasoconstriction; RCVS; reversible; SSRI

Introduction

Reversible cerebral vasoconstriction syndrome (RCVS) is an increasingly recognized acute cerebrovascular condition that may be idiopathic or secondarily provoked.[1,2] It is heralded by an acute, severe headache associated with multifocal cerebrovascular irregularities, which are transient in nature.[1,2] The subsequent clinical course is variable, but may include focal and diffuse neurologic deficits and seizures.[1–3] The clinical manifestations of RCVS-related cerebral infarction or intracranial hemorrhage may be transient or result in permanent deficits.

Intracranial hemorrhage in RCVS is usually subarachnoid, but may be intraparenchymal or even subdural.[4] Intra-parenchymal hemorrhage in RCVS is most often unilateral.[4] We report the first case of a woman with RCVS complicated by bilateral basal ganglia hemorrhages, which was associated with severe clinical deficits that largely resolved, resulting in a near complete clinical recovery.

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Conflict of Interest: None.

Case Report

A 58-year-old previously healthy woman developed a sudden-onset, severe headache. Initial blood pressure was elevated, but subsequent vital signs were normal. She was somnolent and had severe receptive aphasia and moderate bradykinesia.

Head computed tomography (CT) on presentation (day 1) revealed bilateral basal ganglia hemorrhages (Fig 1). Lumbar puncture and toxicology screens were unrevealing. Cerebral MR angiography (MRA) on day 10 showed relatively normal middle cerebral arteries (MCAs) and anterior cerebral arteries (ACAs) (Fig 2A), and MRI at that time showed no evidence of infarction (not shown). However, conventional cerebral angiography on day 11 revealed subtle and multifocal arterial vasoconstriction (Fig 3). Subsequent brain magnetic resonance (MR) imaging on day 14 showed small left MCA infarctions (Fig 4). Repeated MRA on day 21 revealed evolving vascular abnormalities, with both ACAs and MCAs having become severely constricted (Fig 2B). Near resolution of the diffuse intracerebral vasculature constriction on MRA was demonstrated on day 26 (Fig 2C).

She was diagnosed with RCVS, attributed to a selective serotonin reuptake inhibitor (SSRI) prescribed several months prior. The SSRI was stopped at presentation and she was given oral nimodipine. Apart from a minimal receptive aphasia, she made a near full neurologic recovery by hospital day 30 and was discharged home.

Discussion

This case represents a relatively extreme example of RCVS in that the patient was left with a paucity of clinical deficits, despite the development of cerebral infarction and large bilateral basal ganglia intraparenchymal hemorrhages. While unilateral basal ganglia hemorrhages have been reported previously,[4] this represents the first known case in which RCVS has presented with bilateral basal ganglia hemorrhages.

Subarachnoid hemorrhage and multifocal intracranial vascular stenoses are the most common radiologic findings, while intraparenchymal hemorrhages are considerably less common.[1–4] Although subarachnoid hemorrhage (SAH) is relatively common in RCVS, intraparenchymal hemorrhage occurs in only about 10% of patients.[3,4] Older age, female gender, and a history of migraine may be risk factors for RCVS-related intracranial hemorrhage.[4] Other radiologic and clinical features in RCVS may overlap with those seen in reversible posterior leukoencephalopathy.[2,3]

Current evidence suggests that headache and hemorrhage typically precede ischemic complications,[4] as in the case presented here. While the exact mechanisms of hemorrhage in RVCS remain incompletely understood, the current leading theory posits that the initial pathologic process consists of abrupt transient vascular segmental constriction and dilation involving mainly very small vessels, which are frequently angiographically silent.[3] This early process leads to the initial headache and rupture of small vessels.[4] In a second, later stage, major intracranial vessels become involved, also reversibly, resulting in ischemic infarctions. The temporal dynamics of our case is consistent with this hypothesized two-part pathophysiologic process, with early thunderclap headache and bilateral hemorrhage,

followed weeks later by progressive diffuse narrowing of major intracranial arteries and development of ischemic strokes, followed by gradual resolution of the vascular abnormalities. RCVS may occur as an idiopathic process, but is secondary to a defined provocative state or exposure in 60% of cases–vasoactive substances and the postpartum state are common culprits.[2] The presumed provocative factor in the case patient was an SSRI. Given the relationship to exogenous substances and current lack of definitive treatment, identification of RCVS allows the clinician to identify a possible provocative factor and remove or discontinue it.

References

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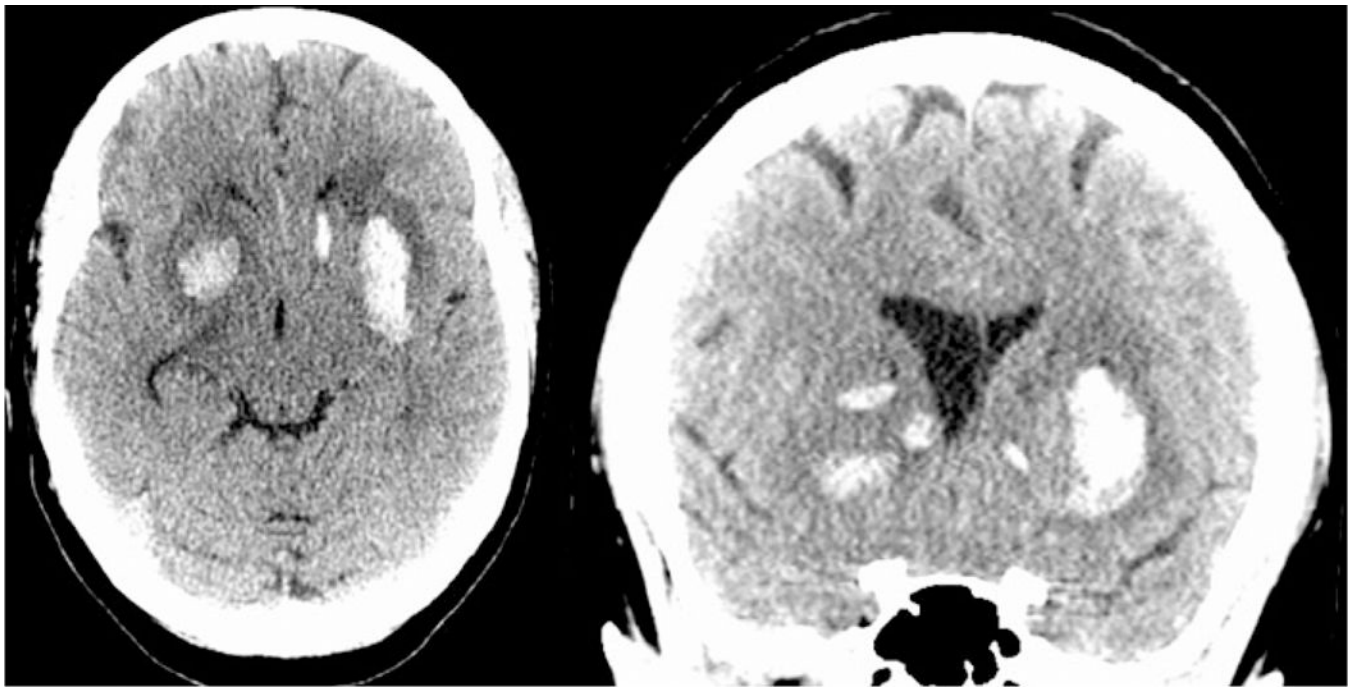

Fig 1.
Head CT on initial presentation (day 1), axial section and coronal reformat, showing bilateral basal ganglia hemorrhages.

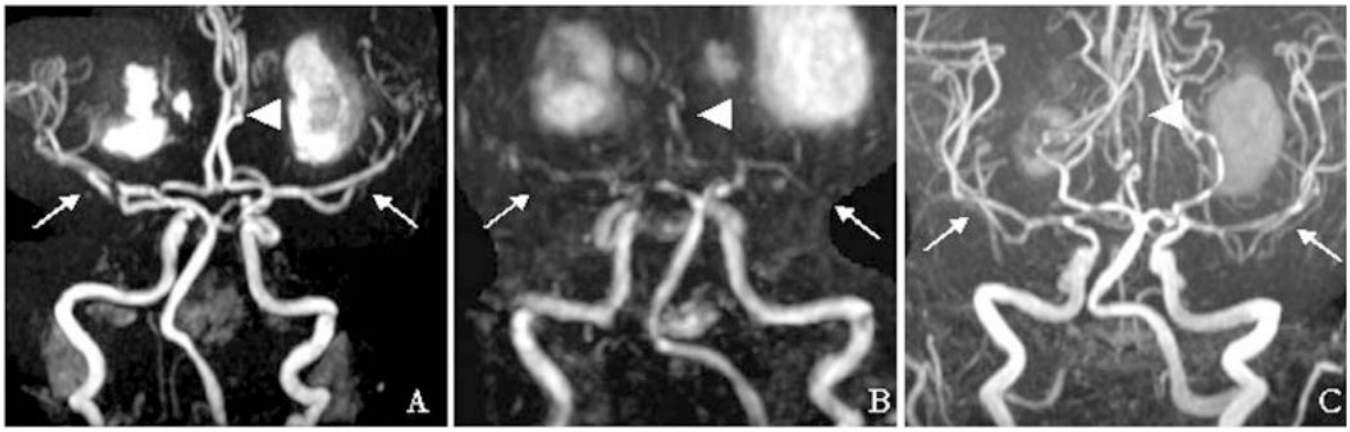

Fig 2.

Sequential head MRA showing evolution of multifocal vascular stenoses. Relatively normal MCAs (arrows) and anterior cerebral arteries (ACAs) (arrowheads) on day 10 are shown in A. Both ACAs and MCAs were severely constricted on day 21 (B). On day 26, there was near resolution of the diffuse intracerebral vasculature constriction (C).

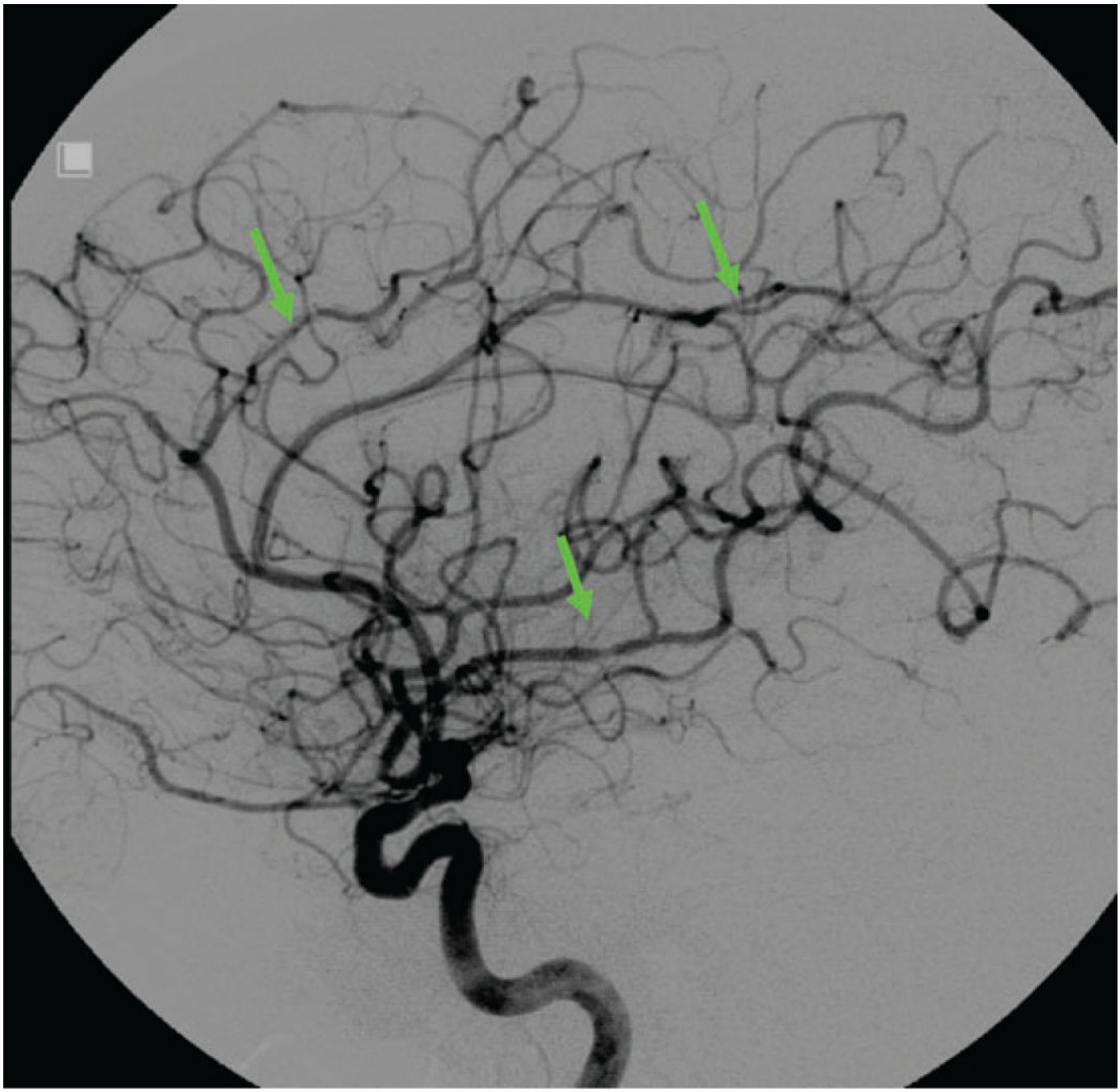

Fig 3.

Conventional cerebral angiography on day 11, right internal artery injection, lateral projection, showing multifocal arterial vasoconstriction (arrows show three examples, but a diffuse process is demonstrated).

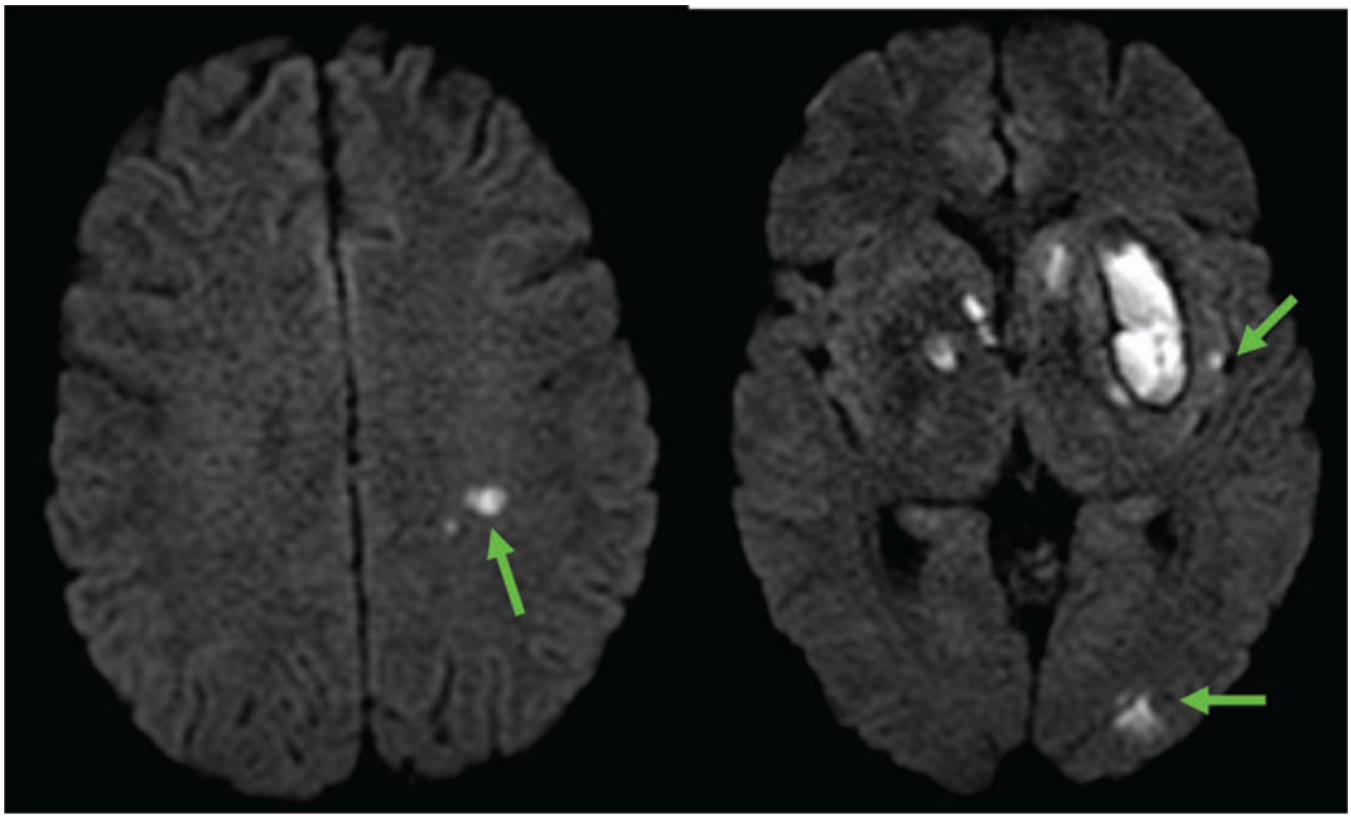

Fig 4.

Head MRI on day 14, diffusion-weighted imaging, showing scattered regions of reduced diffusivity, representing small infarctions (arrows) in the territory of the left middle cerebral artery. On the right panel, basal ganglia hemorrhages are also demonstrated.