Isolated Extracranial Vasospasm and Intracranial Dissection: an Unusual Imaging Manifestation of Reversible Cerebral Vasocclusion Syndrome

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Abstract

Reversible cerebral vasoconstriction syndrome (RCVS) is a distinct clinical syndrome characterized by thunderclap headache and reversible cerebral vasospasm. An association between RCVS and arterial dissection has recently been uncovered [1], whereas vasoconstriction in RCVS is diffuse, bilateral, and intracranial, associated dissection typically involves the cervical vertebral arteries in almost all cases [1]. We report a highly unusual presentation of RCVS, namely, reversible spasm confined to the cervical segments of bilateral vertebral arteries and isolated intracranial dissection, an association hitherto never described.

Introduction

Reversible cerebral vasoconstriction syndrome (RCVS) is a curious clinicoradiologic entity defined by thunderclap headache and reversible vasospasm. An association between RCVS and cervical arterial dissection has recently been uncovered [1], whereas vasoconstriction in RCVS is diffuse, bilateral, and intracranial, associated dissection typically involves the cervical vertebral arteries in almost all cases [1]. We report a highly unusual presentation of RCVS, namely, reversible spasm confined to the cervical segments of bilateral vertebral arteries and isolated intracranial dissection, an association hitherto never described.

Case Report

A 48-year-old woman came to our hospital with thunderclap headache associated with diplopia, vertigo, and dizziness. She had a history of chronic, bi-temporal, headaches for which she occasionally used triptans. She described her present symptoms as being worse in severity, occipital in location, and different in character from her usual headache. Neurologic exam revealed left eye esotropia, left abducens, and left facial nerve palsy. Computed tomography (CT) done in the emergency department was negative for acute intracranial hemorrhage, but showed suspicious hypodensity in the brainstem and left cerebellar hemisphere. An magnetic resonance imaging (MRI) was ordered to further characterize the CT findings, and small acute infarcts were seen in the brainstem and the left inferior cerebellar hemisphere. The basilar artery and the V4 segment of the left vertebral artery showed lack of flow voids and T1 hyperintensity. The cervical segments of the vertebral and carotid arteries were normal. Clinicoangiographic findings were deemed to be consistent with vertebralbasilar dissection, and antiplatelet therapy was initiated accordingly. No specific history of vigorous neck movement, manipulation, or injury was obtained. She had no vascular risk factors and her stroke work up including her coagulation panel, and echocardiogram was unremarkable. Four weeks later, she presented with acute onset of speech difficulty, jaw clenching, malaise, and slumping over in her chair, which lasted 2 min. A repeat MRI of the brain showed no new intracranial findings. The MR angiogram of the neck, however, showed diffuse beadling of bilateral the cervical segments (V2) of bilateral vertebral arteries, which was not seen on the prior angiogram.

An association between arterial dissection and RCVS has been described [1]. Extracranial arterial involvement in RCVS, although rare, has also been reported [2]. Based on this knowledge and given that the cervical segments of the vertebral arteries were normal at initial presentation, a provisional diagnosis of reversible vasocostriction of the extracranial vertebral arteries associated with intracranial arterial dissection was suggested. A follow-up MR angiogram performed about ten weeks
after the first episode showed normal appearance of bilateral V2 segments with resolution of beaded vasoconstriction, thereby confirming the diagnosis. The patient has had no recurrence of headache or other neurological symptoms, continues to be on antiplatelet medication and regular clinical follow-up.

Discussion

RCVS is a distinct neurologic entity characterized clinically by thunderclap headache and radiologically by reversible and segmental cerebral arterial vasospasm. Affected individuals typically fall in the 20–50-year age group and show a female preponderance. Two-thirds of the cases of RCVS have an identifiable trigger, commonest being post-partum state, and use of vasoactive
drugs (including triptans and serotonin reuptake inhibitors). Alterations in cerebral vascular tone leading to cerebral vasoconstriction are believed to result in RCVS, although the underlying pathophysiologic mechanism is not well understood [3]. Hyperacute onset of severe headache that peaks within 60 s (thunderclap) is the classical presenting symptom; headache may occur in isolation or be accompanied by nausea, vomiting, diplopia, photosensitivity, and blood pressure elevation. Migraine is a frequent association, although patients with RCVS describe the symptoms as being different from their usual headaches [3]. The clinical picture can be complicated by the presence focal neurologic deficits, seizures, cortical subarachnoid or intraparenchymal hemorrhage, cerebral infarction, and concomitant posterior reversible encephalopathy [4]. Given the symptomatology and the acuity of onset, most patients are encountered in the emergent setting with aneurysmal rupture and cerebral venous thrombosis being the commonest clinical differentials. However, cerebrospinal fluid analysis, head CT, and initial angiogram are often unremarkable. The imaging hallmark of RCVS is reversible beading of medium-to-large arteries with multifocal segments of narrowing interspersed with normal-caliber vessels. Arterial involvement typically starts at the level of the dural penetration with involvement of the extracranial segments being rare [5]. Digital subtraction angiography is considered the gold standard, but is invasive and is, thus, often replaced by MR angiography, especially for the purpose of documenting resolution of vasoconstriction within three months [6,7]. Radiological findings may lag the clinical presentation and are often not seen until a week after symptom onset [3]. Ducros and Bowser [3] suggested that the delay in the detection of vasoconstriction on imaging may be due to segmental spasm beginning in the small peripheral arterioles and progressing centripetally toward the larger vessels.

An association between RCVS and cervical arterial dissection has been described in recent years; the largest study was published by Mawet et al. [1], comprising of 20 patients where dissection of the vertebral arteries was seen in 19 patients. This is in contrast to isolated dissection, which involves the carotid arteries majority of the cases [8]. In addition to the usual clinical presentation, neck pain was seen in 75% of patients in this cohort [1]. The frequency of symptomatic infarcts was, however, lower than isolated cervical artery dissection (5% versus 79%) [1]. Vasoconstriction was diffuse, bilateral, and was limited to the intracranial segments. The chronology and cause effect relation between RCVS and dissection was not been well established. Of the 20 cases described, dissection was diagnosed before in 3 patients, at the time in 10 patients, and after RCVS in 7 patients. A high index of clinical suspicion and sequential vascular imaging is, thus, required to uncover the association between RCVS and dissection.

Our case was challenging because dissection was limited to the intracranial vertebral and the basilar arteries and

Figure 3. (a) Coronal images from an MR angiogram of the neck done after two weeks of initial onset shows beaded appearance of the cervical segments of the vertebral arteries. (b) MR angiogram obtained at ten weeks following the first onset of symptoms shows resolution of vasoconstriction and normalization of vertebral arteries.
the neck vasculature was initially normal—a finding not seen in any of the 20 patients described by Mawet et al. [1]. Thunderclap headache at presentation and a history of vasoactive drug usage were the only clues in our patient that prompted a follow-up angiogram. The other unusual finding was that vasoconstriction confined to the cervical/V2 segments of the vertebral arteries with sparing of the intracranial vasculature—again not seen in any of the patients in the largest series. Resolution of vasoconstriction within three months, however, helped us confirm this elusive diagnosis. An association between intracranial dissection and extracranial vasoconstriction limited to the vertebral arteries as seen in our patient has never been reported. We believe that this could at least in part be secondary to lack of routine inclusion of the neck vessels in the follow-up of potential cases, given our continuously increasing knowledge about the entire spectrum of RCVS. Whether this was an isolated phenomenon or occurs frequently is uncertain and studies with larger patients would be needed to characterize neck vessel involvement in RCVS.

Management of RCVS includes intensive unit level care, withdrawal of any suspicious triggers, including vasoactive medications, symptom relief, blood-pressure control, and seizure prophylaxis [3]. Nimodipine has been shown to provide symptomatic relief [3]. Most patients with RCVS have good prognosis and symptoms typically resolve in three weeks [3]. Patients with vertebral artery dissection and associated RCVS also have a good overall outcome and a lower incidence of symptomatic infarcts.

References