

Vessel Wall MRI to Differentiate Between Reversible Cerebral Vasoconstriction Syndrome and Central Nervous System Vasculitis

Preliminary Results

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Background and Purpose—Prospective differentiation between reversible cerebral vasoconstriction syndrome and central nervous system vasculitis can be challenging. We hypothesized that high-resolution vessel wall MRI would demonstrate arterial wall enhancement in central nervous system vasculitis but not in reversible cerebral vasoconstriction syndrome.

Methods—We identified all patients with multifocal segmental narrowing of large intracranial arteries who had high-resolution vessel wall MRI and follow-up angiography at our institute over a 4-year period and performed a detailed chart review.

Results—Three patients lacked arterial wall enhancement, and these all had reversal of arterial narrowing within 3 months. Four patients demonstrated arterial wall enhancement, and these had persistent or progressive arterial narrowing at a median follow-up of 17 months (range, 6–36 months) with final diagnoses of central nervous system vasculitis (3) and cocaine vasculopathy (1).

Conclusions—Preliminary results suggest that high-resolution contrast-enhanced vessel wall MRI may enable differentiation between reversible cerebral vasoconstriction syndrome and central nervous system vasculitis. (*Stroke*. 2012;43:860-862.)

Key Words: magnetic resonance imaging ■ reversible cerebral vasoconstriction syndrome ■ vasculitis ■ vasoconstriction ■ vessel wall

Proposed diagnostic criteria for “reversible cerebral vasoconstriction syndrome” (RCVS) are sudden-onset severe headache, multifocal segmental narrowing of cerebral arteries, lack of aneurysmal subarachnoid hemorrhage, near-normal cerebrospinal fluid, and spontaneous resolution of arterial narrowing within 3 months.^{1,2} Early discrimination between RCVS, and its principal differential, central nervous system (CNS) vasculitis, is important: RCVS is treated with observation or possibly calcium channel blockers, whereas CNS vasculitis is treated with steroids and immunosuppression.¹ However, the diagnosis of RCVS is currently confirmed only in retrospect, when arterial narrowing resolves.

High-resolution vessel wall MRI is an emerging technique for characterizing intracranial arterial disease.^{3,4} Large artery CNS vasculitis is associated with arterial wall thickening and enhancement.^{4,5} RCVS is a disorder of arterial tone regulation, and the limited histopathologic data available suggest an absence of arterial wall inflam-

mation^{6,7}; therefore, we hypothesized that patients with RCVS would lack arterial wall enhancement. We reviewed our database of intracranial vessel wall MRI to identify all patients with multifocal segmental narrowing of large intracranial arteries and angiographic follow-up and report the findings.

Patients and Methods

Patients

We routinely perform vessel wall MRI for patients with intracranial arterial narrowing of unclear etiology. From a database of 114 patients studied between January 2006 and December 2010, we identified all patients with multifocal segmental narrowing of large intracranial arteries, vessel wall MRI performed at presentation, and angiographic follow-up. Multifocal narrowing was defined as multiple smooth concentric regions of narrowing that are long relative to the diameter of the artery, excluding the focal irregular stenoses that are typical of atherosclerosis.⁸ We performed a detailed chart review for each case. Institutional Review Board approval was obtained.

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Table. Results

Subject No.	Age, y	Sex	High-Resolution Vessel Wall MRI		Angiographic Degree of Narrowing	Management	Angiographic Follow-Up		Final Diagnosis* (Evidence)
			Arterial Wall Thickening	Arterial Wall Enhancement			Length, mo	Outcome	
1	32	F	Yes	Absent/minimal	Severe	Calcium channel blocker and steroid	3	Resolution	RCVS†
2	56	F	Yes	Absent/minimal	Moderate	Calcium channel blocker	1	Resolution	RCVS†
3	45	F	Yes	Absent/minimal	Moderate	Calcium channel blocker and steroid	1.5	Resolution	Possible RCVS‡
4	69	F	Yes	Yes	Severe	Antiplatelet agent and steroid	24	Persistence	Giant cell arteritis (superficial temporal artery biopsy)
5	53	F	Yes	Yes	Severe	None	6	Persistence	CVS vasculitis (biopsy-proven sarcoidosis)
6	19	M	Yes	Yes	Moderate	Steroid and immunosuppressant	6	Persistence	Probable ¹⁰ CNS vasculitis (clinical course, angiography, CSF)
7	19	F	Yes	Yes	Severe	None	36	Persistence	Cocaine vasculopathy (symptom onset immediately after first-time cocaine use)

F indicates female; M, male; CVS, cerebrovascular; CNS, central nervous system; CSF, cerebrospinal fluid.

*In none of the cases were the clinical or imaging features suggestive of other causes of intracranial arterial stenosis such as dissection or fibromuscular dysplasia.

†Fulfilled the proposed diagnostic criteria for reversible cerebral vasoconstriction syndrome.^{2,3}

‡Transient arterial narrowing that did not fulfill all diagnostic criteria for reversible cerebral vasoconstriction syndrome due to a cerebrospinal fluid leukocytosis.

Vessel Wall MRI Protocol

MRI was performed using a Signa HDx 3.0-T scanner with an 8-channel head coil (GE Healthcare, Milwaukee, WI). Vessel wall protocol included a time-of-flight MR angiography of the circle of Willis, T1-weighted black blood vessel wall sequence (single inversion recovery-prepared 2-dimensional fast spin echo acquisition with field of view=22×22 cm, acquired matrix=512×512; slice thickness=2 or 3 mm; total slab thickness 2–3 cm, TR/TI/TE=2263/860/13 ms) before and after intravenous gadolinium (with constant scan parameters) and T2-weighted vessel wall sequences, which are not included in this Brief Report. Images were obtained through the most severely narrowed arterial segments in each patient. Each vessel wall sequence was performed in both the short and long axes of the artery of interest.

Image Interpretation

A neuroradiologist, blinded to vessel wall imaging and clinical data, graded the angiographic arterial narrowing as mild (<50%), moderate (50%–79%), or severe (>80%) for each study. A different neuroradiologist, blinded to angiography and clinical data, categorized vessel wall thickness as “normal” (barely perceptible) versus “thickened,” and comparing the pre- and postgadolinium images, classified arterial wall enhancement in narrowed segments as “absent/minimal” versus “present” for each study. Categorization was qualitative based on clinical experience interpreting a wide range of intracranial vessel wall MRI examinations.

Results

Seven patients satisfied the inclusion criteria. Median age was 45 years (range, 19–69 years) and 6 of 7 patients were women. Presenting symptoms were acute headache (n=2), neurological deficit (n=2), and headache with deficit (n=3). Laboratory testing included a serum vasculitis work-up in 7 of 7 patients and cerebrospinal fluid analysis in 5 of 7

patients. High-resolution vessel wall MRI findings, patient management, angiographic follow-up, and final diagnoses are listed in the Table. Representative images are provided in Figures 1 and 2.

Discussion

We have used high-resolution contrast-enhanced vessel wall MRI to characterize the intracranial arterial wall in

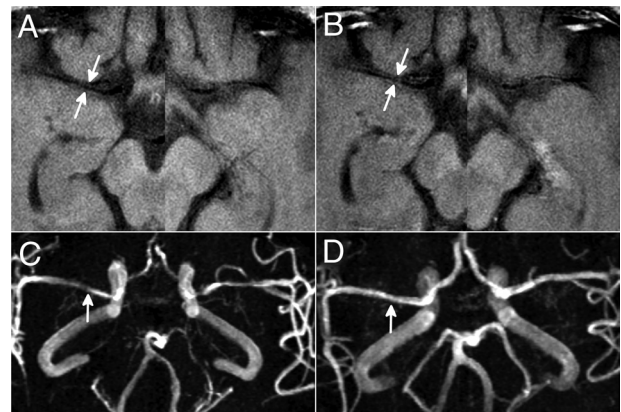


Figure 1. Reversible cerebral vasoconstriction syndrome. Axial T1-weighted vessel wall MRI pre- (A) and post- (B) gadolinium at presentation demonstrate arterial wall thickening (arrows) but absent/minimal arterial wall enhancement. Note the slice positioning is different for the right and left middle cerebral artery images. MR angiography at presentation (C) shows segmental narrowing of the anterior, middle (arrow), and posterior cerebral arteries bilaterally. Three-month follow-up angiogram (D) shows resolution of arterial narrowing.

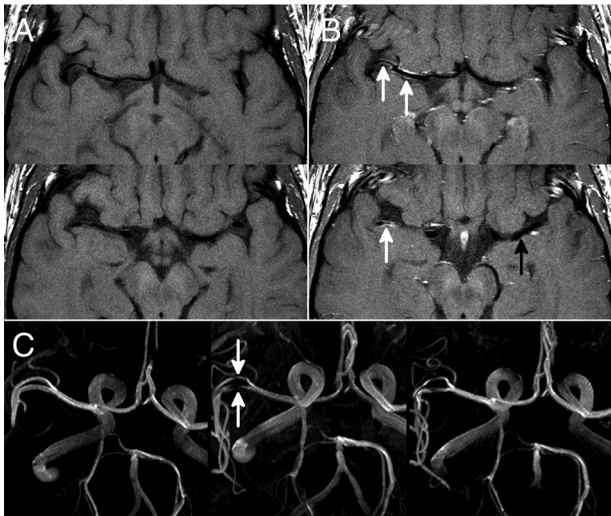


Figure 2. Central nervous system vasculitis. Axial T1-weighted vessel wall MRI pre- (A) and post- (B) gadolinium at presentation demonstrate arterial wall thickening and enhancement (arrows). MR angiogram at the time of vessel wall imaging (middle image in C) shows narrowing of the right middle cerebral artery (arrows) compared with a MR angiogram obtained 2 months earlier (left image in C). Follow-up angiogram 3 months after vessel wall MRI (right image in C) shows persistent narrowing.

patients with multifocal segmental narrowing of large intracranial arteries.

In patients with RCVS, vessel wall MRI demonstrated arterial wall thickening and a lack of arterial wall enhancement. This is consistent with the pathology of transient vasoconstriction. In vasospastic arteries, smooth muscle cells shorten in length with increased overlap among cells resulting in nearly a 500% increase in wall thickness for a 60% luminal narrowing.⁹ Lack of arterial wall enhancement is concordant with the limited histopathologic data in RCVS showing an absence of arterial wall inflammation.^{6,7} The 1 patient with transient arterial narrowing who did not fulfill all diagnostic criteria for RCVS was taking a selective serotonin reuptake inhibitor, a known risk factor for RCVS. Vessel wall MRI findings in this patient were identical to those of the patients who satisfied all criteria for RCVS.

In patients with persistent arterial narrowing, diagnoses were CNS vasculitis (N=3) or cocaine vasculopathy (N=1), and vessel wall MRI demonstrated circumferential arterial wall thickening and enhancement. Like RCVS, cocaine vasculopathy is characterized by vasoconstriction, but unlike RCVS, cocaine vasculopathy demonstrates arterial wall in-

flammation on histopathologic evaluation.¹¹ Two of the patients with persistent arterial narrowing were included in previous studies^{4,11} of vessel wall MRI.

This preliminary study is limited by the small sample size. Although we used the existing diagnostic criteria for RCVS, it is possible that some cases truly represent variants of “vasculitis” with early resolution of arterial narrowing. Also, patients in this study received differing treatments, potentially affecting vessel wall enhancement and clinical course. Of the 2 patients who received steroids before vessel wall imaging, 1 had a final diagnosis of RCVS and the other CNS vasculitis.

Conclusions

Our preliminary results raise the possibility of using high-resolution contrast-enhanced vessel wall MRI to differentiate between vasoconstriction and CNS vasculitis.

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Disclosures

None.

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Abstract

可逆性脳血管攣縮症候群と脳血管炎を鑑別する血管壁 MRI 所見 — 予備段階の結果

Vessel Wall MRI to Differentiate Between Reversible Cerebral Vasoconstriction Syndrome and Central Nervous System Vasculitis — Preliminary Results

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背景および目的：可逆性脳血管攣縮症候群と脳血管炎の前向きな鑑別は困難である。高解像度の血管壁 MRI では、脳血管炎の動脈壁の増強が示されるが可逆性脳血管攣縮症候群の動脈壁の増強は示されないとの仮説を立てた。

方法：4年間に当施設で高解像度の血管壁 MRI とそれについて血管造影を受け、頭蓋内主幹動脈の多巣性の分節状狭小化がみられたすべての患者を同定し、詳細なカルテの評価を行った。

結果：3例の患者では動脈壁の増強はみられず、これらの患者全員に3カ月以内に動脈狭小化の逆転が生じた。4例の患者に動脈壁の増強がみられ、これらの患者は中央値17カ月(範囲6～36カ月)時の追跡調査で持続性または進行性の動脈狭小化が認められ、最終診断は脳血管炎(3例)とコカイン血管症(1例)であった。

結論：予備段階の結果から、高解像度の造影血管壁 MRI により、可逆性脳血管攣縮症候群と脳血管炎を鑑別できる可能性が示唆される。

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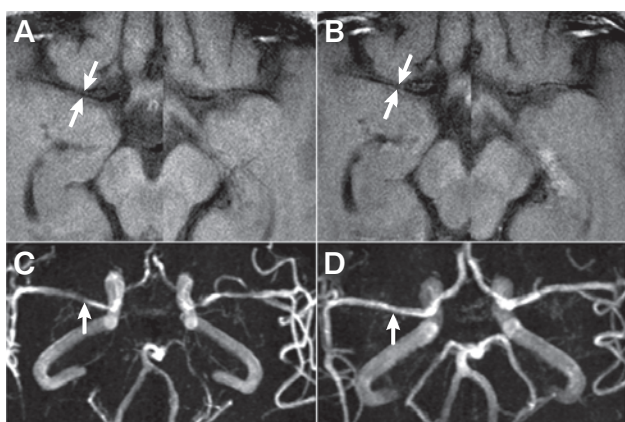


図 1

可逆性脳血管攣縮症候群。診察時のガドリニウム投与前(A)および投与後(B)の軸断 T1 強調血管壁 MRI では動脈壁の肥厚(矢印)が認められるが、動脈壁の増強はほぼ/まったくみられない。左右の中大脳動脈像ではスライスの位置が異なることに注意。診察時の MR 血管造影(C)では、前、中(矢印)、および後大脳動脈の両側性の分節状狭小化が認められる。3カ月の追跡調査時の MR 血管造影(D)では、動脈狭小化の消失が認められる。

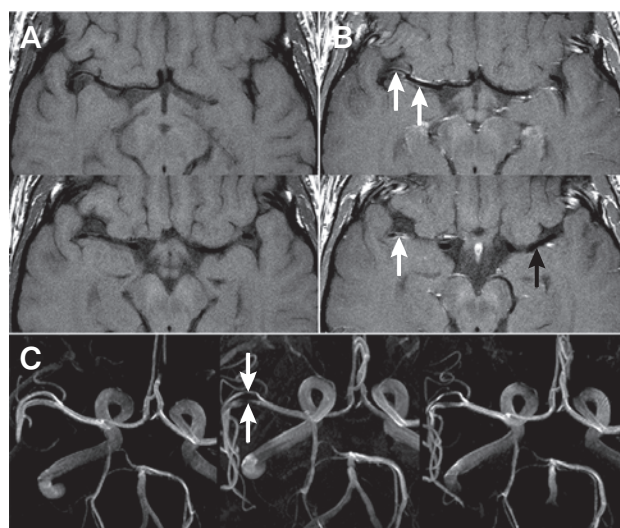


図 2

脳血管炎。診察時のガドリニウム投与前(A)および投与後(B)の軸断 T1 強調血管壁 MRI では、動脈壁の肥厚および増強(矢印)が認められる。血管壁画像診断時の MR 血管造影(Cの中央の像)では、2カ月前に撮影された MR 血管造影(Cの左の像)と比較して、右中大脳動脈の狭小化(矢印)が認められる。血管壁 MRI から3カ月後の追跡調査の MR 血管造影(Cの右の像)では、狭小化の持続が認められる。

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