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CASE REPORT

Imaging Characteristics of Reversible Cerebral Vasoconstriction Syndrome: an Under-recognised Cause of Severe Headache

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ABSTRACT

Reversible cerebral vasoconstriction syndrome is an under-diagnosed disease that every radiologist should know about. This report demonstrates two female patients who presented with severe headache with subsequent angiographic findings of 'bead and string' appearances of the cerebral arteries at Circle of Willis, which resolved spontaneously within 3 months. Diagnosis of reversible cerebral vasoconstriction syndrome was made. The clinical and imaging characteristics in different modalities are discussed.

Key Words: *Cerebral arterial diseases; Headache disorders, primary; Magnetic resonance angiography; Vasoconstriction; Vasospasm, intracranial*

中文摘要

可逆性腦血管收縮綜合徵的影像學特徵：劇烈頭痛的一種尚未充分認識的病因

李嘉樂、蕭容媛、AT Ahuja

可逆性腦血管收縮綜合徵是一個常常未被明確診斷的疾病，對此放射科醫生都應有所了解。本文報告兩名有劇烈頭痛的女性病人，血管造影顯示其威利斯（Willis）環腦動脈出現串珠狀表現，但於三個月內自行緩解，後被確診為可逆性腦血管收縮綜合徵。本文討論此病的臨床及影像學特徵。

INTRODUCTION

Reversible cerebral vasoconstriction syndrome (RCVS) constitutes an under-recognised but clinically important diagnosis, because it can be complicated by a cerebrovascular accident. The syndrome is often misdiagnosed as it resembles primary angiitis of the central nervous system, aneurysmal subarachnoid haemorrhage, cerebral venous sinus thrombosis and arterial dissection. Demonstration of a 'bead and string pattern' of segmental vasoconstriction of the cerebral arteries in a patient presenting with severe headache, and with complete or near-complete reversibility of vasoconstriction variably within 3 months is the most specific imaging characteristic of this disorder. In order to diagnose the condition earlier, radiologists should be aware of the clinical and imaging characteristics of RCVS. The two cases described here illustrate the imaging characteristics of RCVS in different modalities, including computed tomography (CT), magnetic resonance imaging (MRI), and digital subtraction angiography (DSA).

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CASE REPORTS

Case 1

A 49-year-old woman presented with sudden-onset severe frontal throbbing headache in July 2009. She had vomited once before the admission. The patient could recall 2 similar episodes before, which resolved spontaneously. There were no other focal neurological symptoms or signs. Lumbar puncture (LP) revealed no abnormalities. Blood autoimmune markers were all normal.

Initial non-contrast CT of the brain showed no abnormality. Cerebral magnetic resonance angiography (MRA) revealed no aneurysm. The cerebral magnetic resonance venogram (MRV) revealed absence of venous thrombosis. Computed tomography angiography (CTA) of the cerebral circulation was undertaken a few days later owing to worsening persistent headache and a 'string and bead' appearance with multiple segmental stenoses involving essentially all the cerebral vessels in both the anterior and posterior cerebral circulation bilaterally (Figure 1a). The major differential diagnoses were cerebral vasculitis or RCVS.

The symptoms improved gradually following treatment with calcium channel blockers. Cerebral CTA repeated 1 month later showed partial resolution of the multiple segmental stenoses (Figure 1b). Follow-up cerebral CTA 3 months later showed complete resolution of the beading appearance of the cerebral arteries (Figure 1c). The final diagnosis was RCVS.

Case 2

A 42-year-old woman presented with severe occipital headache and neck pain for 5 days in December 2008. A LP was unremarkable. An initial CT brain (at a private hospital) showed no abnormality. Moreover, MRI including MRA/MRV showed no aneurysm or dural sinus thromboses, but with high T2 signal intensity bilaterally at the anterior aspects of the basal ganglia (Figure 2a). A provisional diagnosis of cervicogenic headache was entertained coupled with non-specific T2 signal intensity bilaterally in the anterior parts of the basal ganglia. The patient was then discharged.

One week later the patient presented to hospital again with complex partial seizure and persistent occipital headache. CT and MRI brain showed white matter oedema bilaterally in the occipital and parietal lobes (Figures 2b and c). MRA was again unremarkable (Figure 2d). A few days after admission, she developed increasingly severe thunderclap headaches and CT and MRI brain then revealed a new infarct in the right middle cerebral artery territory and the previously noted bilateral occipital and parietal lobe white matter oedema had resolved (Figures 2e and f). The cerebral MRA showed beading appearance of the cerebral arteries at the level of Circle of Willis (Figure 2g). DSA of the right internal carotid artery and its branches confirmed the multiple constrictions and dilatations in the anterior and middle cerebral arteries (Figure 2h).

The patient was treated conservatively with calcium channel blockers. The follow-up cerebral MRA and DSA after 2 months showed resolution of the vasoconstrictions (Figures 2i and j). The overall features were compatible with RCVS. A history obtained retrospectively revealed that the patient had taken oral contraceptive pills before the headaches.

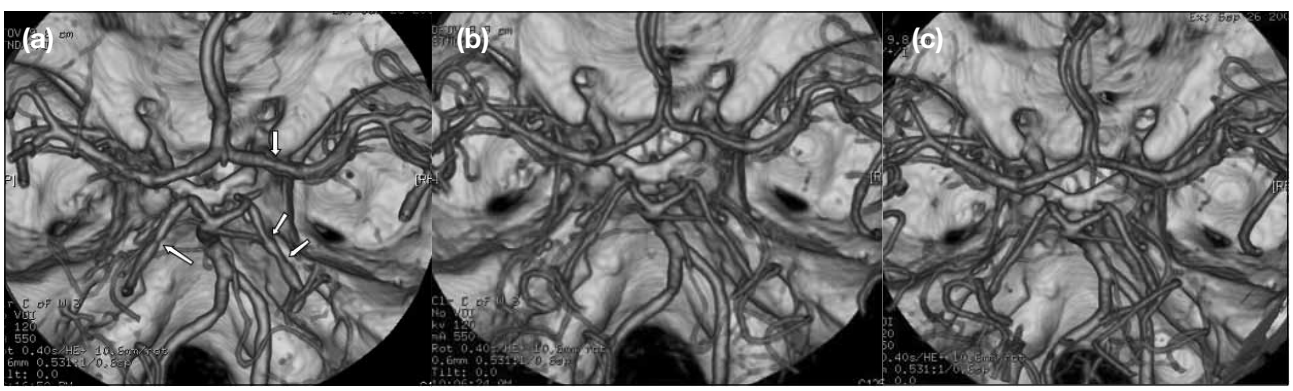

Figure 1. Case 1: (a) Cerebral computed tomography angiography (CTA) on admission shows beading of the anterior and posterior cerebral arteries at Circle of Willis with multiple stenoses and dilatation (arrows). (b) Repeated cerebral CTA 1 month later shows partial resolution of the beading. (c) Cerebral CTA 3 months later shows complete resolution of the segmental vasoconstrictions.

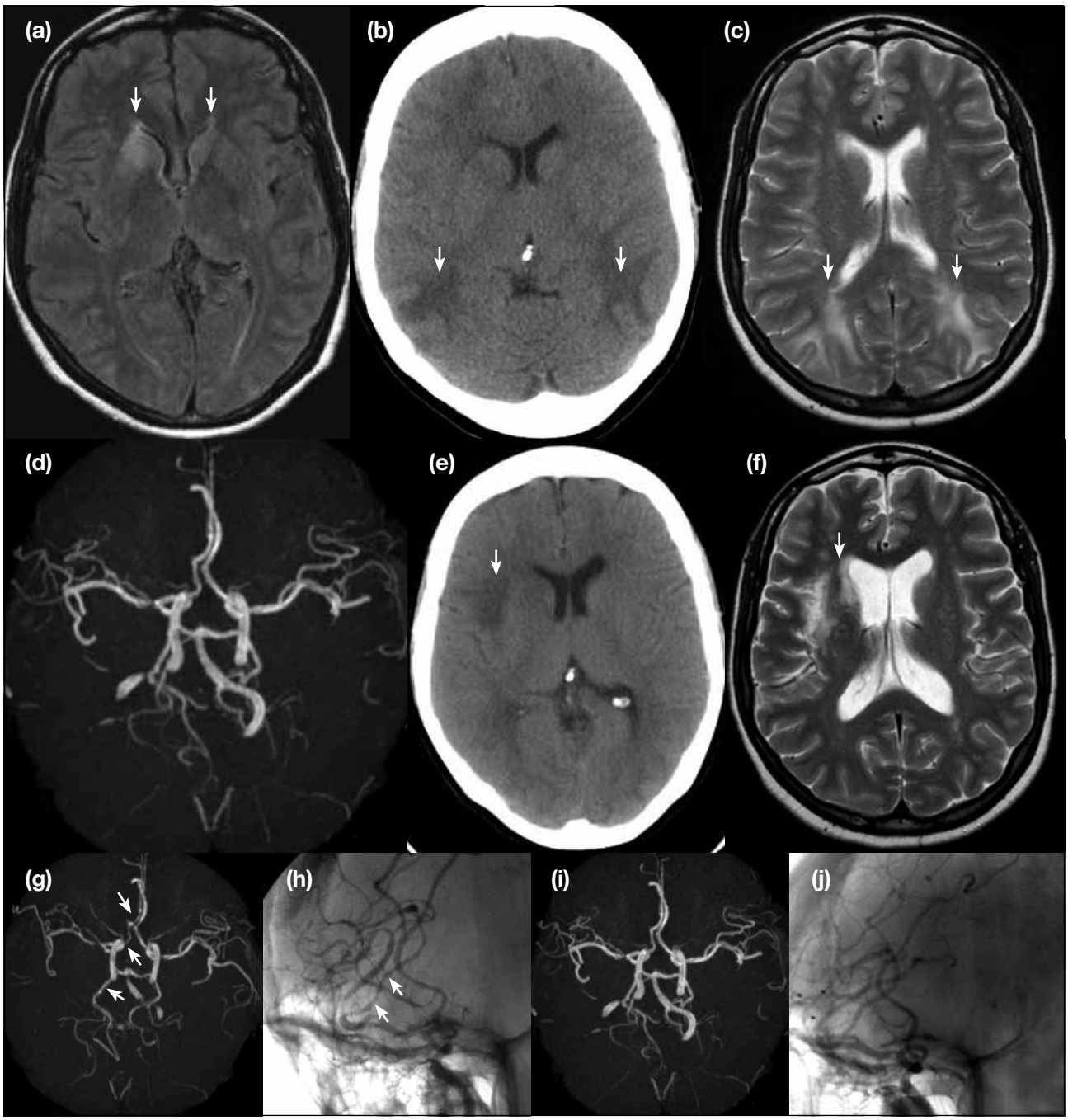

Figure 2. Case 2: (a) The initial axial T2-weighted fluid-attenuated inversion recovery (FLAIR) magnetic resonance imaging (MRI) brain shows non-specific mild T2 hyperintensities bilaterally at the anterior basal ganglia (white arrows). (b) The axial non-contrast computed tomography (CT) brain shows white matter hypodensity at the bilateral occipitoparietal regions (white arrows). (c) The axial T2-weighted FLAIR MRI brain was consistent with white matter oedema at the same regions (white arrows). (d) The first cerebral magnetic resonance angiography (MRA) was unremarkable. (e) The axial non-contrast CT brain shows hypodensity at the right insula involving the basal ganglion, which was suspicious of an acute right middle cerebral artery infarct (white arrow). (f) The subsequent axial T2-weighted MRI brain suggests acute cerebral infarct at the region of insula along the right middle cerebral territory and resolution of the T2 hyperintensities at the bilateral occipitoparietal regions (white arrow). (g) The cerebral MRA shows new-onset 'bead and string' appearance of the anterior and posterior cerebral arteries (arrows) bilaterally. (h) The cerebral angiogram of the right internal carotid artery and its branches confirms multiple beading appearance of the anterior and middle cerebral arteries (arrows). (i) The cerebral MRA shows resolution of the multiple focal cerebral arterial vasoconstrictions after 2 months. (j) Subsequent digital subtraction angiography of the right internal carotid artery and its branches show resolution of the vasoconstrictions after 2 months.

DISCUSSION

RCVS is a new unifying term that was previously referred to as the Call syndrome (or the Call-Fleming syndrome), benign acute cerebral angiopathy or migrainous vasospasm. It occurs primarily in females.[1,2]

It is characterised by acute, severe, recurrent headache and a 'bead and string' appearance of the cerebral arteries which resolves spontaneously in 1 to 3 months.[3-5] It may or may not be associated with neurological symptoms or signs. One study suggested that recurrent thunderclap headache lasting a few days to 2 weeks is the clinical hallmark of RCVS,[6] and was also present in our 2 cases.

Multiple precipitating factors have been described, including the peripartum period, bathing, exposure to drugs or Chinese herbal medicines, alcohol, medications and blood products, catecholamine-secreting tumour and vasoactive substances.[6-8] The oral contraceptive pill may have been the precipitating factor in our second case. However, often there is no clear precipitant.[2]

Multiple segmental vascular narrowing involving the cerebral artery at Circle of Willis and its immediate branches resulting in 'bead and string' angiography is the characteristic imaging appearance evident within minutes to weeks of onset. Large- and medium-sized cerebral arteries are predominantly involved. The most specific imaging evidence for RCVS depends on the timely demonstration of complete or near-complete reversibility of the vasoconstriction, which invariably ensues within 3 months.[9]

Cross-sectional imaging (including CT and MRI) is the investigation of choice to diagnose RCVS. More invasive imaging techniques (such as DSA) are supplementary tools for the diagnosis of equivocal or difficult cases. Research has revealed that RCVS is associated with severely elevated blood velocities demonstrable by transcranial Doppler, which may therefore be an ideal imaging tool for the follow-up of patients as it entails no radiation.[10]

The major differential diagnosis includes primary angiitis of the central nervous system, aneurysmal subarachnoid haemorrhage, cerebral venous sinus thrombosis and arterial dissection.[7,11] LP should be performed to exclude 'CT-negative' subarachnoid haemorrhage and inflammatory conditions, such as infection and cerebral vasculitis. Unruptured aneurysm is an uncommon incidental finding.[12] Complicated haemorrhage should be excluded by imaging or even LP. MRA/MRV or CTA/CT venography should be carried out to rule out dural venous thrombosis or arterial dissection.

CT and MRI are also useful for detecting stroke (ischaemic or haemorrhagic), which is an important major complication of RCVS. The stroke predominantly affects the border zone[13-15] and is likely due to the vasoconstriction as demonstrated in case 2. In addition, reversible T2-weighted white matter hyperintensities are quite commonly shown in imaging studies, as in case 2. These findings are also probably due to vasoconstriction-induced ischaemia. Uncommon complications such as subarachnoid haemorrhage may also be noted in the imaging study and confused with the primary subarachnoid haemorrhage with secondary vasoconstriction, in which case the time sequence and the results of LP are important differentiators.[13]

Treatment options include: observation, calcium-channel blockers (nimodipine and verapamil), and possibly high-dose corticosteroids. Induced hypertension or blood pressure lowering and rarely endovascular therapy[9] are other less commonly used options. Overall, the neurological outcome is good and most patients recover fully with conservative treatment.[13]

RCVS is an under-diagnosed and under-recognised disorder and commonly misdiagnosed as one of its simulators.[11] Radiologists should know this entity well, and consider the diagnosis in all patients presenting with severe thunderclap headaches with typical beading appearance of the cerebral arteries.[4,5]

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