Case Report

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A Case of Central Nervous System Vasculitis Mimicking Reversible Cerebral Vasoconstriction Syndrome in Young Stroke: A Diagnostic Conundrum

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ABSTRACT

Central Nervous System Vasculitis (CNSV) is a rare cause of stroke in young adults and may closely mimic Reversible Cerebral Vasoconstriction Syndrome (RCVS), a more common non inflammatory vasculopathy. Distinguishing between these entities is crucial because their treatment and prognosis differ. The authors report a 40-year-old female who presented with one month of progressive right-sided hemicranial headache followed by onset of left hemiparesis. Initial non contrast Computed Tomography (CT) brain showed right parietal infarct with haemorrhagic transformation. Magnetic Resonance Imaging (MRI) with diffusion-weighted imaging revealed multiple watershed infarcts in the right fronto-parietal region, while gradient echo sequences demonstrated right parieto-occipital lobar haemorrhage with overlying convexity subarachnoid haemorrhage. Time-of-flight MR angiography showed multifocal segmental narrowing with irregular contour of the right M2 segment of the Middle Cerebral Artery (MCA), favouring a working diagnosis of RCVS, and she was started on nimodipine. However, Cerebrospinal Fluid (CSF) analysis showed mild pleocytosis with elevated protein, and inflammatory work-up revealed raised acute phase reactants with positive Antinuclear Antibody (ANA) and anti-U1 Ribonucleoprotein (RNP), suggestive of an underlying Mixed Connective Tissue Disease (MCTD)-associated CNSV. She was treated with high-dose intravenous corticosteroids followed by monthly intravenous cyclophosphamide pulses, with good clinical and radiological improvement. The present case is unique because multifocal vasculitis involvement with combined ischaemic watershed infarcts and concomitant lobar and convexal haemorrhage simulated RCVS in a young woman without systemic autoimmune features. The report underscores the need to reconsider a vasculitis aetiology when atypical imaging, abnormal CSF and positive autoimmune markers coexist in a presumed case of RCVS, in order to avoid misdiagnosis and to initiate timely immunosuppression.

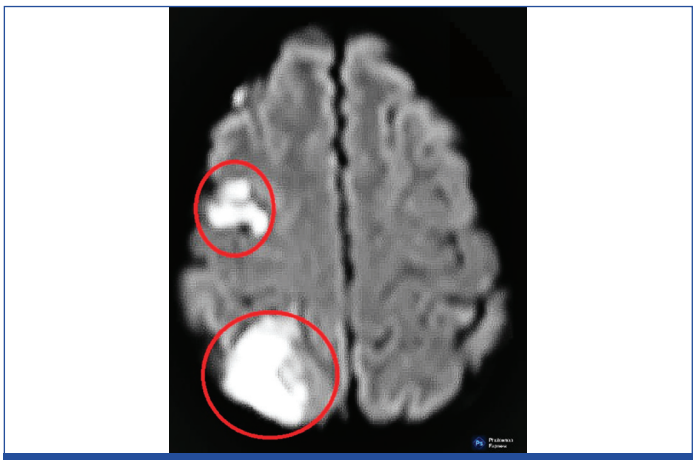
Keywords: Brain ischaemia, Connective tissue disease, Hemiplegia, Intracranial haemorrhages, Middle-aged

CASE REPORT

A 40-year-old female presented with one month history of progressive episodic headache which was holocranial, non radiating and occasionally associated with photophobia and nausea. There was no history of vomiting, fever, seizures or altered sensorium. Her past medical history was unremarkable; she had no prior history of any systemic co-morbidities or any diagnosed autoimmune or connective tissue disorder. There were no recent pregnancy, puerperium, head trauma or recent infections. On admission, she was found to have hypertension, with a Blood Pressure (BP) of 160/100 mmHg. While other vitals were stable. The general examination was unremarkable. There were no focal deficits on neurological examinations at admission.

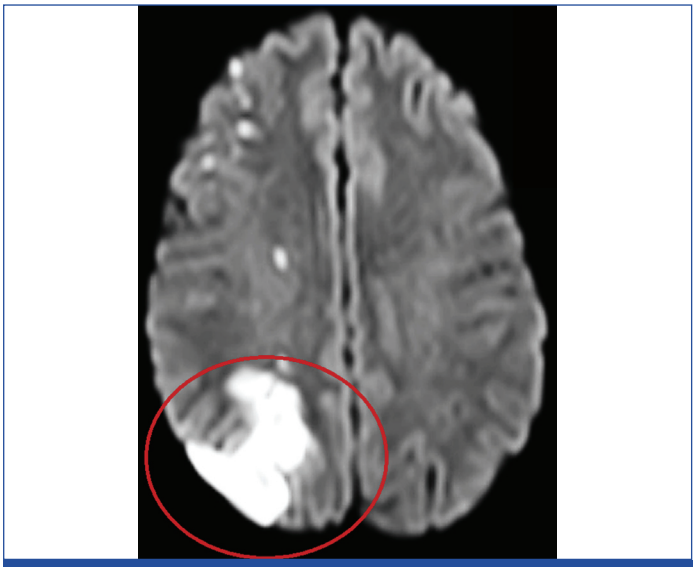
She had Computed Tomography (CT) brain done outside before admission and the images revealed an acute infarct in the right occipital region. Based on the CT findings, and the history she was initially treated as a case of ischaemic stroke and newly diagnosed hypertensive with antiplatelets (tab. Aspirin 75 mg and Clopidogrel 75 mg) and statins (Tab. Atorvastatin 40 mg). She underwent an extensive evaluation for the aetiology of young-onset stroke.

On day 3 of the hospital stay, her headache worsened which was progressive along with sudden onset left hemiplegia and left upper motor neuron facial palsy. Repeat CT brain showed an acute infarct in the right MCA territory. MRI of brain showed acute watershed infarcts in two regions, first one in the right Anterior Cerebral Artery (ACA) and MCA and the second in the MCA with Posterior Cerebral Artery (PCA) territory [Table/Fig-1,2]. There were areas of blooming in the right parieto-occipital cortex on Susceptibility Weighted Imaging (SWI) suggestive of haemorrhagic foci, with a small Sub Arachnoid Haemorrhage (SAH) in the right parietal sulcus [Table/Fig-3,4]. Magnetic Resonance Angiography (MRA) showed multisegmental narrowing with irregular contour of the right M2 segment of MCA [Table/Fig-5].

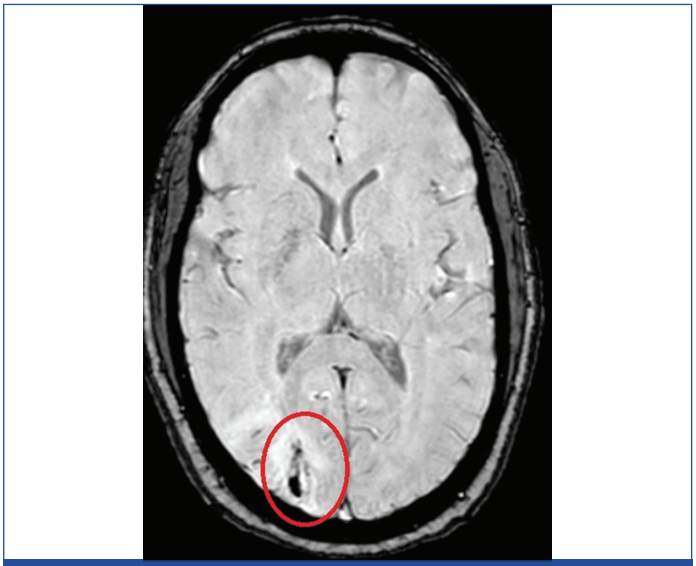


[Table/Fig-1]: Axial DWI image shows diffusion restriction in the right parietal region, corresponding to watershed territory between MCA and PCA. Areas of diffusion restriction are also noted in the right centrum semiovale, corresponding to watershed territory between ACA and MCA.

At this point, with a presentation of subacute onset of recurrent worsening headaches followed by acute infarcts at this young age, the differential diagnosis sought with CNS restricted vasculitis and RCVS. These findings of multiple watershed infarcts, presence of minimal SAH in the right parietal sulcus, no intracranial carotid narrowing, female sex, appropriate age group, in the clinical background of recurrent headaches, favoured a diagnosis of RCVS.



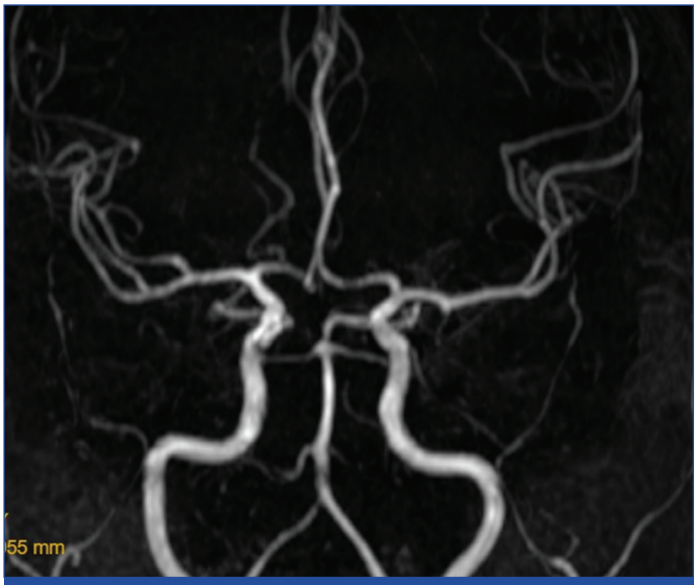
[Table/Fig-2]: Axial Diffusion-weighted Imaging (DWI) image shows larger area of diffusion restriction s/o acute infarct in the right parietal cortex and scattered smaller areas of infarcts in right frontal cortex (Red circle).



[Table/Fig-3]: An axial Gradient Echo (GRE) image shows blooming in the right parieto-occipital cortex, suggestive of parenchymal haemorrhage (Red circle).

[Table/Fig-4]: An axial GRE image shows blooming in the sulcus of right parietal region, suggestive of subarachnoid haemorrhage (Red circle).

Later CSF analysis was sent and the resulted with normal findings with presence of five cells (all lymphocytes), glucose was 85 mg/dL, and protein 45 mg/dL, further favoured RCVS. However, the possibility of medium vessel vasculitis could not be ruled out. So, she started on Nimodipine 60 mg sixth hourly for RCVS and vasculitis work up was sent immediately. Various diagnosis possibilities thought



[Table/Fig-5]: MRA showing multisegmental narrowing with irregular contour of the right M2 segment of MCA.

of include Primary Angiitis of the CNS (PACNS), CNS lupus, Anti-Neutrophil Cytoplasmic Antibodies (ANCAs) associated vasculitis, Antiphospholipid Antibody (APA) syndrome, polyarteritis nodosa, CNS tuberculosis, Neuro Behcet's and Deficiency of Adenosine Deaminase-2 (ADA2) syndrome.

To establish a provisional diagnosis from the differential diagnosis, the patient was subjected to a comprehensive series of laboratory tests. Complete Blood Count (CBC) showed microcytic hypochromic anaemia (haemoglobin 7 g/dL), and the rest of the reports were within normal limits. Erythrocyte Sedimentation Rate (ESR) was 104 mm and C-Reactive Protein (CRP) was elevated (26 mg/dL). Urine routine was normal. Electrocardiogram (ECG) and Echocardiography (ECHO) were normal. ANA was strongly positive (3+) with speckled nuclear pattern. Clinically, the patient did not fulfill criteria for Systemic Lupus Erythematosus (SLE) later due to the laboratory work-up, SLE diagnosis also sought. Complement levels were normal. Cytoplasmic and perinuclear ANCA (cANCA and pANCA) were negative, hence ANCA-associated vasculitis was ruled out. APA was done (anticardiolipin, lupus anticoagulant and beta-2 glycoprotein antibody) and found to be negative, further ruled out APA syndrome.

Extractable nuclear antigen immunoblot was strongly positive for U1-RNP and Ro52 antibodies signifies the presence of MCTD. Although U1-RNP was positive, the patient did not fulfill other clinical criteria for MCTD, hence it was ruled out. Schirmer's test was done to rule out Sjogren's, and was normal and there were no other clinical features of the same. She had no other organ or skin involvement consistent with any of the systemic vasculitis or Behcet's syndrome. Tests sent for ADA2 deficiency resulted negative. Brain biopsy was not feasible in the present setting.

After the initiation of nimodipine (30 mg Q4 hrly) for possible RCVS, no clinical improvement was noted. In view of rapid neurological worsening and positive antinuclear antibodies, CNS vasculitis was suspected. She was therefore given pulse methylprednisolone 1 g once daily for three days followed by oral prednisolone 60 mg once daily. Nimodipine was stopped. After one week of hospital stay her power improved from 0/5 to 4/5 in lower limb and 0/5 to 1/5 in upper limb. Hydroxychloroquine (HCQ 100 mg twice daily) was added at discharge to maintain the remission in autoimmune CNS vasculitis and to reduce the further thrombotic risk.

During follow-up for a week, after confirmation of CNS vasculitis as the diagnosis, she was started on monthly cyclophosphamide at the dose of 0.75 g/m^2 (1gm/month). Totally, she received six doses of cyclophosphamide and steroids (oral prednisolone 60 mg once daily) were tapered during this period. The strength in her affected

limbs is now fully restored to 5/5 after three months of follow-up, and she currently has no complaints. Then the patient was advised regular follow-up with neurology for monitoring disease activity and to monitor long-term immunosuppression and any autoimmune evolution.

DISCUSSION

The authors report a case of 40-year-old female who initially presented with recurrent headaches and was found to have acute infarcts. Imaging showed watershed infarcts, subarachnoid haemorrhage, and multifocal arterial narrowing, suggestive of RCVS. However, autoimmune markers and a lack of response to nimodipine shifted the diagnosis towards CNS vasculitis. The patient showed remarkable improvement following immunosuppressive therapy. The present case highlights the diagnostic complexity in differentiating CNS vasculitis from RCVS.

Vasculitis refers to the inflammation of the blood vessel walls, which can result in vessel narrowing, occlusion, or rupture, ultimately leading to tissue ischaemia or haemorrhage. Although systemic vasculitis is uncommon, CNS restricted vasculitis is even rarer and represents a challenging diagnosis, particularly in young adults presenting with stroke [1,2]. PACNS, a subtype of CNS vasculitis, is a rare but important cause of stroke in younger populations, accounting for less than 1% of all strokes and often presenting with non-specific neurological symptoms such as headache, cognitive dysfunction, or seizures [1,3,4]. RCVS, in contrast, is characterised by transient, non inflammatory arterial narrowing typically triggered by vasoactive substances or postpartum state [5,6]. Theoretically, both may exhibit multifocal arterial narrowing and headaches, RCVS commonly presents with thunderclap headaches and has a benign, self-limiting course [7-9]. In contrast, CNS vasculitis often shows progressive neurological decline and elevated inflammatory markers [7,9,10]. Diagnostic differentiation is aided by CSF analysis, where lymphocytic pleocytosis and elevated protein may favour vasculitis, while normal CSF is typical of RCVS. Vessel wall MRI, though not widely available, is an emerging tool showing arterial wall enhancement in vasculitis but not in RCVS [8].

Moreover, strokes in young adults account for approximately 10-15% of all strokes and presents a broad differential diagnosis distinct from other populations [11,12]. Common aetiologies include cardioembolic, arterial dissections, prothrombotic states, and inflammatory vasculopathies, including CNS-vasculitis and RCVS [11]. Inflammatory causes like PACNS, although rare, must be considered in cryptogenic strokes, especially when imaging reveals multifocal infarcts, haemorrhagic components, or atypical vascular changes [12]. The present patient, a 40-year-old woman with recurrent headaches and watershed infarcts, exemplifies this diagnostic dilemma. The patients evolving neurological deficits and positive autoimmune markers highlighted vasculitis as a rare yet treatable cause of young stroke, emphasising the need for early recognition and tailored work-up in this age group.

Few comparative studies clearly distinguish CNS-vasculitis from RCVS [Table/Fig-6] [6,7,10]. Singhal AB et al., found that recurrent or isolated thunderclap headaches with normal imaging, border-zone infarcts, or vasogenic edema, strongly favoured RCVS and developed the RCVS2 score based on such variables, including female sex, vasoconstrictive triggers and SAH [10]. Based on this scoring system, the patient's initial presentation and normal CSF favoured RCVS by this score. Yet, Singhal AB et al., also noted that eccentric narrowing was more typical of CNS-vasculitis, unlike symmetric, concentric, and smooth taper lesions seen in RCVS [10]. While de Boysson H et al., retrospectively analysed 110 patients with PACNS and 173 patients with RCVS and reported that thunderclap headaches, SAH, and vasogenic edema predominated in RCVS, conversely, persistent motor deficit, seizures, acute ischaemic stroke and Intracerebral Haemorrhage (ICH) were more common in PACNS [7]. The present patient had no thunderclap headache but presented with persistent motor deficit and eccentric MCA narrowing, favouring CNS vasculitis, though some imaging still mimicked RCVS. Notably, cases with SAH and irregular angiography may still be vasculitis as shown by three patients in the study by de Boysson H et al., [7] and similar findings in other studies [6,13].

Features with reference	RCVS	CNS vasculitis	Present case
Headache type [7,10]	Recurrent or isolated thunderclap headaches	Gradual, persistent headaches	No thunderclap headache
CSF findings [10]	Typically, normal	Often shows lymphocytic pleocytosis, elevated protein	Normal CSF
Angiographic pattern [10]	Symmetric, smooth tapering	Eccentric, irregular narrowing	Eccentric MCA narrowing
Common imaging findings [7,10]	Border-zone infarcts, vasogenic oedema, SAH	Multifocal infarcts, ischaemic strokes, ICH	Watershed infarcts, minimal SAH
Clinical course [7,10]	Benign, self-limiting	Progressive neurological decline and elevated inflammatory markers	Progressive
Persistent neurological deficit [7]	Less common	More common	Present
CSF analysis [6,7]	Normal findings	Lymphocytic pleocytosis and elevated protein levels	Lymphocytes (five cells) and increased protein and glucose
Treatment [6,7]	Responds to CCBs, worsens with steroids	Steroids/ immunosuppressants	No response to nimodipine; improved with steroids
Final diagnosis	Less likely	More likely	CNS-vasculitis

[Table/Fig-6]: Comparison of key distinguishing features of RCVS and CNS-vasculitis with the present case [6,7,10].
Ducros A et al., 2010, Paris, France – 89 patients with RCVS [6]; Singhal AB et al., [10] 2016, Multicentre (USA, Turkey, Hong Kong) – 159 patients with RCVS and 47 patients with PACNS; de Boysson H et al., [7] 2018, Multicentre French cohorts – 110 patients with PACNS; 173 patients with RCVS

Preliminary data have emerged advocating for the use of vessel wall MRI in cases where such differentiation may be challenging. In a study of seven patients, the patients with RCVS had arterial wall thickening and a lack of arterial wall enhancement on vessel wall MRI, consistent with the pathophysiology of transient vasoconstriction [8]. As this was not available for the present case, the clinical course of the illness and further work-up guided the treatment plan. Also, presence of U1-RNP antibodies have been associated with vasculitis manifestation, including CNS involvement even in the absence of full-blown MCTD [14]. This serological profile supported the diagnosis of CNS vasculitis and justified the intuition of immunosuppressive therapy, including HCQ as a preventive strategy against future systemic autoimmune evolution.

Treatment strategies for CNS vasculitis and RCVS differ fundamentally due to their distinct pathophysiology. CNS vasculitis necessitates prompt initiation of high-dose corticosteroids, often followed by immunosuppressive agents such as cyclophosphamide or rituximab, to prevent irreversible neurological damage and relapses [1,3,9]. In contrast, RCVS is managed conservatively with calcium channel blockers like nimodipine and the removal of precipitating factors, as corticosteroids are generally avoided as they worsen the RCVS [5,9,10]. In the present patient, initial assumption of RCVS led to nimodipine therapy without clinical improvement which yielded further differential diagnosis. The subsequent rise in inflammatory markers and positive ANA and antibody against U1-RNP prompted steroid initiation and finally, a dramatic response to pulse methylprednisolone and complete subsequent resolution with cyclophosphamide clinched the diagnosis of CNS vasculitis, resulting

in rapid neurological recovery. Moreover, hydroxychloroquine was added at discharge in order to maintain the remission in autoimmune CNS vasculitis and to reduce the further thrombotic risk as suggested by various literatures [15].

CONCLUSION(S)

The present case highlights the diagnostic challenge in differentiating CNS-vasculitis from RCVS in young stroke patients. Despite overlapping clinical and radiological features, timely recognition of elevated inflammatory markers and autoimmune serological findings guided appropriate therapy. The patient's dramatic response to immunosuppression emphasised the importance of early intervention. Such rare presentations necessitate a high index of suspicion and comprehensive evaluation. Prompt diagnosis can significantly alter outcomes in otherwise disabling conditions.

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