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RAPIDLY PROGRESSIVE PRIMARY CENTRAL NERVOUS SYSTEM VASCULITIS: A RARE CASE WITH MULTIFOCAL SMALL AND MEDIUM VESSEL INVOLVEMENT

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ABSTRACT

Introduction: Primary central nervous system vasculitis (primary CNS vasculitis) is a rare inflammatory disorder that affects small-to-medium-sized cerebral vessels, often leading to recurrent strokes. Diagnosis is vague due to non-specific neurological symptoms. Imaging findings, cerebrospinal fluid (CSF) analysis and exclusion of systemic vasculitis are essential for diagnosis. Early immunosuppressive therapy is crucial, but the prognosis remains unpredictable.

Case description: A 45-year-old female with diabetes, hypertension and multiple ischaemic strokes presented with altered consciousness due to hypoglycaemia. Initial workup at a different hospital suggested cerebral vasculitis based on the findings of cerebral angiography. She had negative systemic vasculitis markers and was treated with corticosteroids and rituximab and discharged. On admission to our facility, an MRI of the brain revealed multiple infarcts of varying ages, and a CTA showed arterial beading, suggestive of primary CNS vasculitis. CSF analysis demonstrated elevated protein and IgG without pleocytosis. Despite high-dose corticosteroids and rituximab, she developed progressive neurological deterioration with new infarcts in the vertebrobasilar territory, leading to brainstem dysfunction and brain death.

Conclusion: Primary CNS vasculitis remains a diagnostic and therapeutic challenge because of its diverse presentation and similarities with other vascular conditions. This case emphasises the importance of early recognition, comprehensive diagnostic evaluation and aggressive treatment to prevent irreversible neurological decline. Disease progression can be rapid even with treatment, highlighting the need for further research to enhance early detection and optimise management strategies.

KEYWORDS

Primary CNS vasculitis, small and medium vessel vasculitis, recurrent strokes, MRI brain abnormalities, immunosuppression

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LEARNING POINTS

- Physicians should be alert in diagnosing primary CNS vasculitis in a young patient with recurrent strokes affecting multiple vascular territories, particularly when systemic vasculitis markers are negative.
- Imaging studies, such as MRI and computed tomography angiography (CTA), play an essential role in diagnosing primary CNS vasculitis, with findings such as arterial beading and a multi-infarct pattern helping differentiate it from atherosclerotic diseases.
- Early recognition and aggressive high-dose corticosteroid and immunosuppressive (rituximab) therapy are essential in primary CNS vasculitis, as delayed treatment can result in rapid neurological deterioration, as seen in this case.

INTRODUCTION/BACKGROUND

Vasculitis encompasses various inflammatory conditions that affect the blood vessels, including primary central nervous system vasculitis (primary CNS vasculitis), a rare condition characterised by inflammation of the brain's vasculature, and less frequently, involvement of the spinal cord[1,2]. It presents with a wide range of symptoms, from transient ischaemic attacks to ischaemic infarction, making diagnosis challenging[2].

Primary CNS vasculitis is uncommon, with an incidence of 2.4 cases per million, primarily affecting middle-aged men[3,4]. Diagnosis is made according to Calabrese's criteria after excluding systemic vasculitis and identifying central nervous system vasculitis through imaging or biopsy[1,4]. Despite breakthroughs in diagnostic methods, brain biopsy is still the gold standard[1,4].

Symptoms include seizures, headaches and stroke-like features, and cerebrospinal fluid (CSF) analysis and elevated inflammatory markers are not definitive[2,3]. For better outcomes, early treatment with corticosteroids and immunosuppressive drugs, such as rituximab, is crucial[4].

Here, we present an unusual case of primary CNS vasculitis, highlighting the challenges in diagnosing and treating this condition.

CASE DESCRIPTION

A 45-year-old Asian female with controlled type 2 diabetes mellitus, hypertension and recurrent ischaemic strokes – the most recent occurring one month prior – presented to the emergency department with a reduced level of consciousness attributed to prolonged hypoglycaemia. Her blood glucose in the ambulance was critically low at 35 mg/dl, necessitating hospital admission. Her baseline functional status included residual left-sided hemiparesis, requiring ambulatory support. She was on multiple medications, including metformin, empagliflozin, gliclazide, mixtard insulin, lisinopril, spironolactone, aspirin, atorvastatin and carvedilol.

In the weeks preceding this presentation, she had been admitted to another hospital after developing left-sided weakness with an accompanying right-sided facial droop. Comprehensive evaluations during that admission – including a computed tomography angiography (CTA) of cerebral vessels – revealed evidence of vasculitic involvement of the middle and small cerebral vessels. Although autoimmune screening was negative, a preliminary diagnosis of primary CNS vasculitis was established, and she was subsequently managed with oral prednisolone and rituximab.

The patient presented to us confused, with spontaneous eye movements, but was non-verbal and unresponsive to commands – Glasgow Coma Scale (GCS) 10/15, E4 V1 M5 – with stable vitals. Physical examination was notable for right-sided gaze deviation, right lower motor neuron facial palsy and reduced muscle tone. Muscle power, as per the Medical Research Council scale, was 1/5 in the left upper and lower limbs, 4/5 in the right upper limb, and 2/5 in the right lower limb, with reduced sensation on the left side compared to the right. Plantar reflexes were upgoing bilaterally. The rest of the systemic examination was unremarkable.

Initial laboratory tests showed leucocytosis – likely steroid-induced – mild anaemia, poorly controlled diabetes and mildly elevated inflammatory markers that were deemed insignificant due to the absence of clinical symptoms. Electrolytes, renal function tests and creatinine levels were normal. An arterial blood gas analysis revealed mild respiratory alkalosis from hyperventilation. Hypoglycaemia, confirmed on repeat testing, was corrected with IV dextrose, partially improving consciousness. The remaining results are summarised in *Table 1*.

Neurology was consulted for suspected new stroke, and the patient was admitted to a high-dependency unit for monitoring. An urgent non-contrast computed tomography (CT) scan of the brain revealed multiple acute, subacute and chronic infarcts across various vascular territories, prompting the resumption of antiplatelets and statins *(Fig. 1)*. An MRI scan of the brain with contrast demonstrated a multi-infarct state, with infarcts of varying ages and new ischaemic changes within the vertebrobasilar territory *(Fig. 2)*. A CTA of the cerebral and neck vessels confirmed primary CNS vasculitis, showing atheromatous changes in the right middle cerebral artery and beading of the right posterior cerebral artery *(Fig. 3)*. As part of the stroke workup, Holter monitoring ruled out arrhythmias, and carotid Doppler ultrasonography identified moderate atherosclerotic changes in both carotid systems without significant stenosis. An interferon-gamma release assay was positive. A chest X-ray was unremarkable, and latent tuberculosis infection was diagnosed. The patient was started on isoniazid 300 mg

daily for six months prior to further immunosuppression.

Based on the clinical presentation, imaging findings and laboratory results, a working diagnosis of ischaemic stroke secondary to primary CNS vasculitis was established. Although initially stable her condition deteriorated, with vitals showing a blood pressure drop to 95/57 mmHg and a GCS of 6/15. She developed cardiac arrest, and a crash call was announced. After eight minutes of cardiopulmonary resuscitation, return of spontaneous circulation was achieved, and she was transferred to the intensive care unit (ICU). A repeat CT scan of the brain revealed new infarcts in the right posterior inferior cerebellar artery *(Fig. 4)*. She received high-dose IV methylprednisolone 1,000 mg daily for five days, as per neurology's plan, followed by a single dose of IV rituximab 1 g, and was maintained on oral prednisone 20 mg daily. In the ICU, her GCS remained 3/15 despite sedation withdrawal, while her blood pressure improved with phenylephrine infusion at 50 mg.

Despite treatment, she developed multiple vertebrobasilar

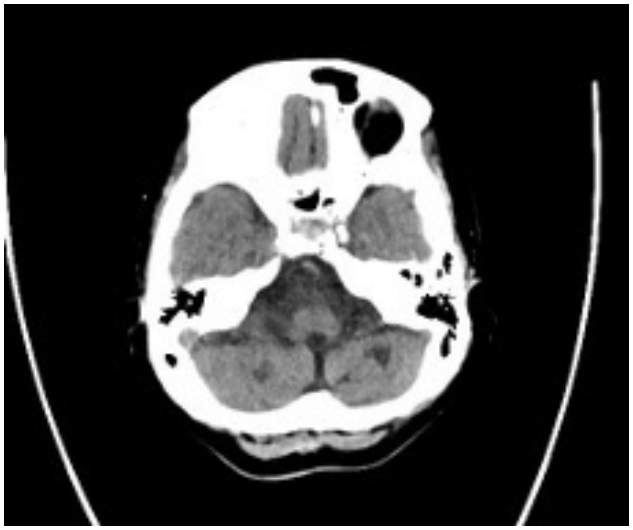


Figure 1. Non-contrast CT scan of the brain showing multiple infarcts of different ages, with both chronic hypodense areas and subtle acute changes.

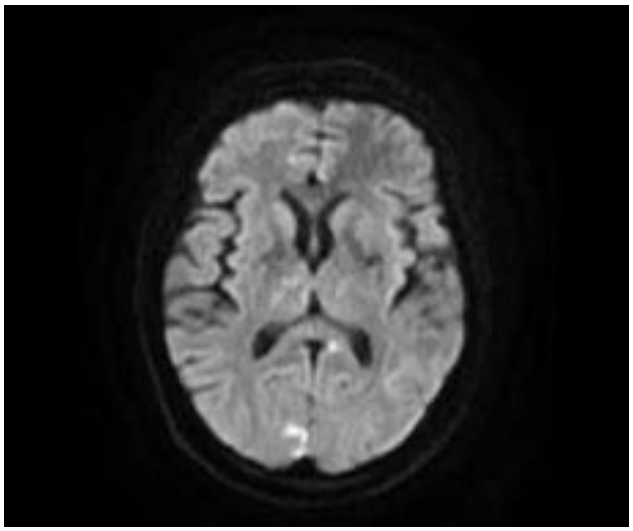


Figure 2. Axial diffusion-weighted MRI demonstrating multiple acute infarcts as hyperintense lesions.

Lab test	Result	Normal range
Full blood count		
WBC (×10³/µl)	21.6	3.6-11
RBC (×10⁶/µl)	4.21	3.6-11
Haemoglobin (g/dl)	11.7	12.0-15
Platelet (×10³/µl)	374	150-410
Neutrophil absolute (×10³/µl)	18.9	2-7
Lactic acid (mmol/l)	1.36	0.5-2.2
HbA1c (%)	11	<5.7
Random blood glucose (mg/dl)	58	64-140
CRP (mg/l)	7.5	<5
Procalcitonin (ng/ml)	0.09	<0.05
Lactic acid (mmol/l)	1.36	0.5-2.2
Urea and electrolytes		
Sodium (mmol/l)	132	136-145
Potassium (mmol/l)	3.4	3.3-4.8
Urea (mg/dl)	35	12-40
Arterial blood gas		
pH	7.47	7.35-7.45
pCO_2 (mmHg)	23.2	40-50
pO_2 (mmHg)	26.2	30-55
HCO_3 (mmol/l)	16.9	21-28
Chest X ray	unremarkable	
Autoimmune work up		
ANA	Negative	Negative
C ANCA (PR3) (AU/ml)	<2	≤19
P ANCA (MPO) (IU/ml)	2	<5
dsDNA Ab (IU/ml)	<10	<10
Rheumatoid factor (U/ml)	<10	0-20
Anti-CCP	Negative	Negative
ESR (mm/hr)	101	<15
C3 (g/l)	1.7	0.8-1.7
C4 (g/l)	0.42	0.15-0.45
CSF		
Protein (mg/dl)	86	15-45
Glucose (mg/dl)	150	40-76
LDH (UI)	48	-
TB PCR	Negative	Negative
AFB smear and culture	Negative	Negative
Culture	Negative	
Meningitis/encephalitis multiplex panel	Negative	

Table 1. Laboratory investigations within the first initial days of admission.

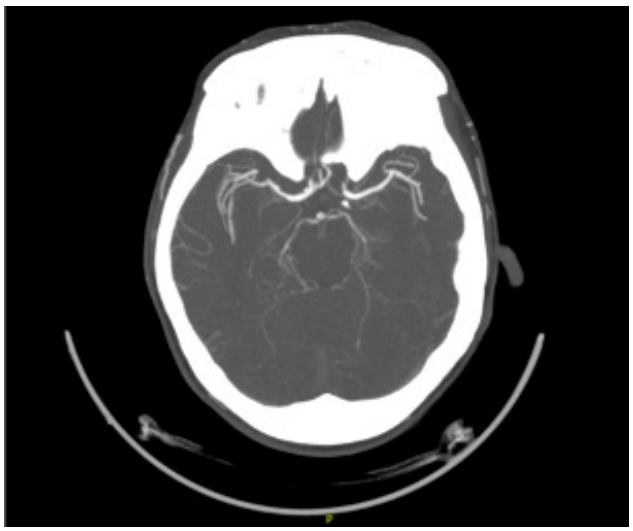


Figure 3. CT angiography showing a beading pattern of alternating arterial narrowing and dilatation, suggestive of cerebral vasculitis.

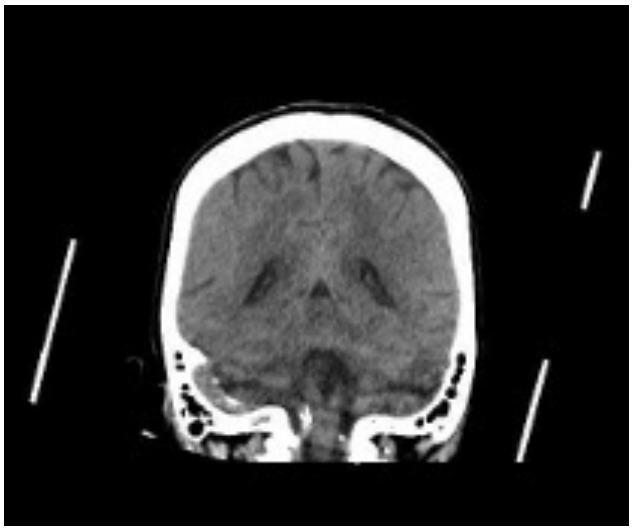


Figure 4. Non-contrast CT scan revealing a new infarct in the posterior inferior cerebellar artery (PICA) territory.

infarcts with neurological findings of fixed, dilated pupils and absent brainstem reflexes. She was declared brain dead after confirmatory tests and transferred for organ donation with family consent.

DISCUSSION

Primary CNS vasculitis is a rare, isolated inflammatory disorder with heterogeneous clinical presentations and an elusive diagnosis. Its non-specific symptoms and absence of systemic involvement often delay diagnosis and treatment. Although the exact pathophysiology is unknown, immune-mediated processes triggered by infections, autoimmunity or other unidentified factors may be involved[1-3]. Imaging modalities, such as MRI and CTA, are critical; arterial beading and multifocal infarcts are hallmark findings. Brain biopsy remains definitive, but it is rarely performed due to its invasiveness and sampling limitations.

Our patient presented with residual left-sided hemiparesis following multiple ischaemic strokes, altered consciousness levels and further neurological deterioration. Imaging studies demonstrated a multi-infarct state with new ischaemic lesions, while a CTA showed arterial beading suggestive of a vasculitic process[2,4]. The diagnosis was confirmed as primary CNS vasculitis due to negative systemic vasculitis markers, negative CSF microbiology and culture, and lack of infectious processes[1,3]; she was treated with high-dose IV methylprednisolone and rituximab[3]. Unfortunately, despite treatment, the patient experienced rapid clinical deterioration, with new infarcts in the vertebrobasilar territory, ultimately leading to brain death[2,4].

Given the aggressive course of primary CNS vasculitis and the need for prompt treatment, as highlighted by the delayed diagnosis in our case, this case contrasts with another primary CNS vasculitis case. This was reported in 2023[5] and related to a 16-year-old female presenting with a history of headaches and epilepsy. The disease followed a more indolent course over 3.5 years, initially being misdiagnosed as a tumour due to its appearance on imaging. Unlike our patient, who experienced multi-infarct strokes, the teenager responded well to early immunosuppression following a biopsy-confirmed diagnosis. These variations in age, comorbidities and disease course highlight the need for more sensitive diagnostic tools and effective therapeutic options.

CONCLUSION

This case highlights the challenges in diagnosing and treating primary CNS vasculitis, particularly in differentiating it from other conditions such as systemic vasculitis. A thorough diagnostic approach, including imaging, CSF analysis and an infectious workup, is essential. Although early aggressive immunotherapy remains the cornerstone of treatment, some cases – such as this one – may follow a rapidly fatal course. Further research is needed to understand the disease mechanisms better and improve treatment strategies.

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