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Case Report

Induced Hypertension in the Management of Primary CNS Vasculitis

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Abstract

In this paper, we report the first documented use of induced hypertension to reverse acute cerebral ischemia in primary CNS vasculitis (PCNSV).

Keywords: Primary Central Nervous System Vasculitis; Induced Hypertension; Ischemic Stroke; Autoimmune Neurology

Introduction

PCNSV is a life-threatening condition characterized by granulomatous inflammation within small cerebral blood vessels [1,2]. The disease is rare, with an estimated annual incidence of 2.4 cases per 1,000,000 person-years, and it affects people of all ages with slight predominance in the seventh decade of life [2,3]. Diagnosis of PCNSV is challenging as it lacks standardized criteria, presents with subtle, nonspecific symptoms, and has a broad differential diagnosis, including intracranial atherosclerosis, reversible cerebral vasoconstriction syndrome, infectious and immune-mediated encephalitis, demyelinating syndromes, and neoplasms [1-8]. Although definitive treatment of PCNSV includes glucocorticoids and cyclophosphamide, days to weeks may be necessary before an effect is observed [1,2,6]. Acute management of PCNSV, particularly the role of induced hypertension to reverse cerebral hypoperfusion, may be necessary as a bridge to immunotherapy but has yet to be highlighted in the literature.

Case Presentation

Our patient is a 74-year-old man who presented with altered mental status. In recent weeks he had developed memory loss and balance problems. Outpatient MR brain showed patchy and confluent T2/FLAIR hyperintensities with diffusion restriction within the deep white matter, right greater than left, that extended to involve the cortex of the right frontal lobe (Figure 1A). In the past week, the patient's functional decline accelerated. He became poorly interactive and was eating and drinking less. He would often not answer questions, and if he did answer, his responses would be incoherent. He would spend most of the day "staring off into space".

On presentation, the subject was awake, alert, and oriented with slow speech and flat affect. Neurologic exam was otherwise nonfocal, and vitals signs, physical exam, and laboratory values were normal. Repeat MR brain showed patchy subacute ischemia in the right frontal lobe and a focus of acute ischemia in the left basal ganglia without contrast enhancement (Figure 1B).

On hospital day (HD) 2, the patient's mentation declined, and he could no longer answer questions or follow commands. A lumbar puncture (LP) was performed with an opening pressure of 11 cmH2O, a white count of 17 cells/microliter with 89% lymphocytes, protein of 59.5 mg/dL and glucose of 72 mg/dL. Extensive CSF and serum evaluation was normal except for positive ANA and anti-centromere B antibodies. EEG showed only mild diffuse slowing without epileptiform discharges or seizures, and echocardiography showed a left ventricular ejection fraction of 65-70% with moderate left ventricular hypertrophy.

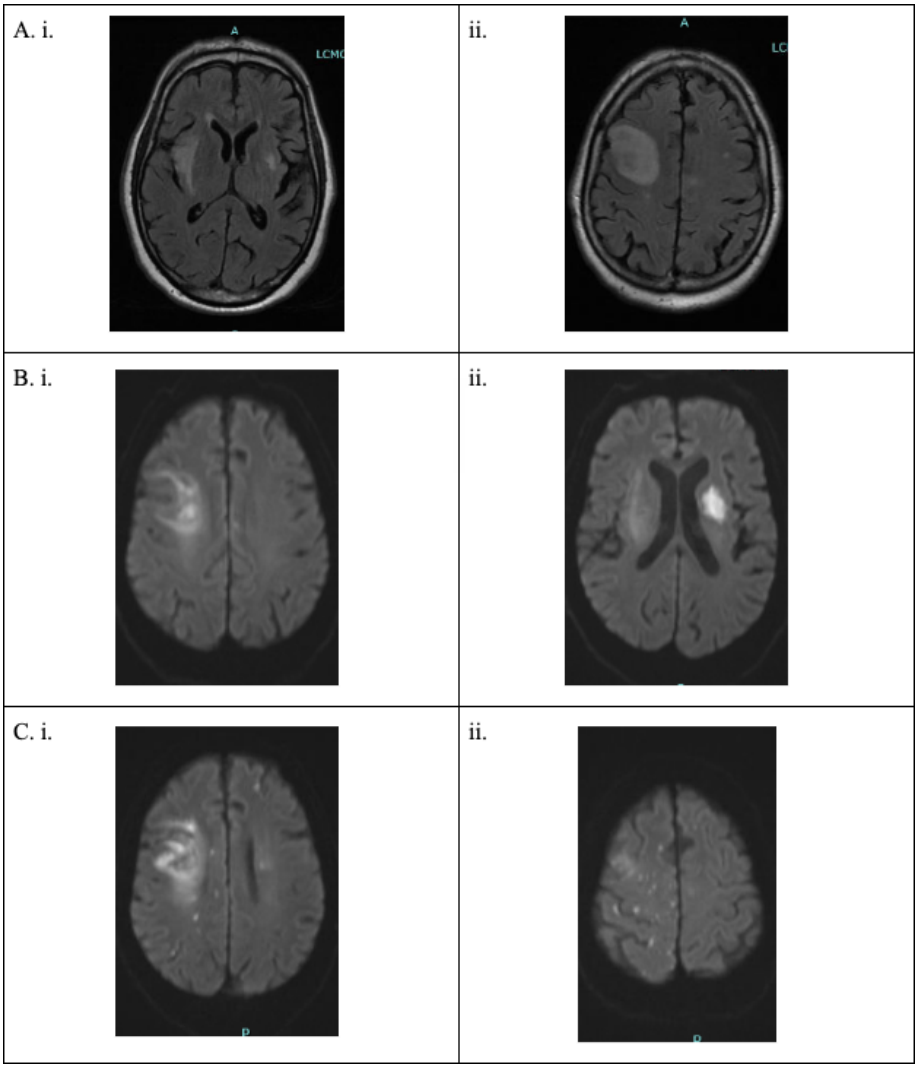

Figure 1: A. Outpatient MRI: i. Axial T2 FLAIR sequence, demonstrating right greater than left white matter and insular changes. ii. Axial T2 FLAIR sequence with well-circumscribed cortical signal change. B. MRI on HD 1: i. Axial DWI sequence with patchy diffusion restriction in the right frontal lobe. ii. Axial DWI sequence with acute ischemia in the left basal ganglia. C. MRI on HD 12: i-ii. Axial DWI sequence demonstrating bilateral punctate foci of acute ischemia.

A digital subtraction angiogram was performed on HD 3. This angiogram showed multifocal intracranial luminal irregularity consistent with either intracranial atherosclerosis or autoimmune or infectious vasculitis. There was complete occlusion of the left A1 and proximal A2 segments, with vascularization of this territory by MCA collaterals (Figure 2A).

The patient was then started on 5 days of pulse methylprednisolone for CNS vasculitis, as well as aspirin and clopidogrel and high-intensity statin. After one day of methylprednisolone, the patient experienced an improvement in awareness, could follow commands, and could feed himself. He was transitioned to prednisone 60 mg daily and continued to show slow if fluctuating improvement in his mental status.

A repeat angiogram was performed on HD 11 which showed interval worsening of luminal narrowing, particularly in the anterior circulation (Figure 2B). Shortly afterward the subject developed left hemineglect and hemiparesis. Repeat MR brain with perfusion showed multiple foci of acute ischemia in bilateral cerebral hemispheres, right greater than left, involving the cortical surface and subcortical white matter without clear perfusion deficits (Figure 1C).

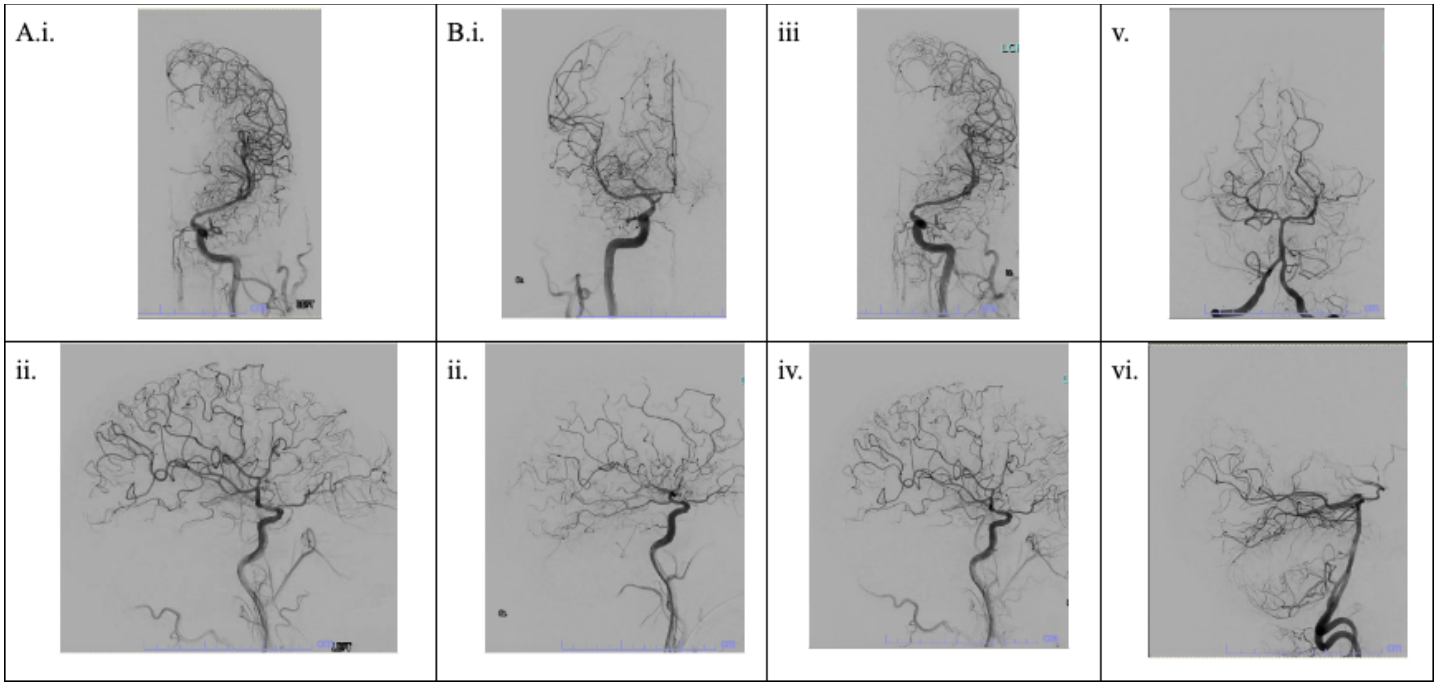

Figure 2: A. Cerebral Angiogram HD 3: AP (i) and lateral (ii) projections of cerebral angiogram demonstrating multifocal luminal narrowing, left A1 occlusion, and revascularization of the ACA territory via MCA collaterals. Overall picture concerning for CNS vasculitis. B. Cerebral Angiogram HD 11: AP (i, iii, v) and lateral (ii, iv, vi) projections of the right anterior circulation (i, ii), left anterior circulation (iii, iv), and posterior circulation (v, vi). Persistent occlusion of the left ACA and worsening of multifocal stenoses consistent with CNS vasculitis.

At this point, our management focused on reversal of cerebral hypoperfusion from PCNSV. Immediately, induced hypertension to a systolic blood pressure (SBP) of 180-220 mmHg was started with norepinephrine. At a higher blood pressure, the patient became more interactive and talkative with less hemineglect and hemiparesis. Rescue angioplasty was contemplated but not performed due to his response to induced hypertension in addition to the severity of his proximal occlusion and extent of distal stenoses.

Two separate attempts to liberalize the SBP goal during the following 10 days led to a decline in exam and recurrent focal deficits (Figure 3). Each recrudescence resolved after increasing the SBP goal and escalating the norepinephrine dose. By HD 21, blood pressure goals were normalized, and he was weaned off vasopressors entirely. On HD 26, a right frontal brain biopsy was consistent with a diagnosis of primary CNS vasculitis with resultant infarct. Cyclophosphamide was started on HD 32, complicated by pancytopenia, cardiomyopathy, and arrhythmia. Consequently, cyclophosphamide was changed to rituximab. The patient was discharged to rehabilitation and is now at home, where he ambulates with assistance with mild impairments in speech and cognition.

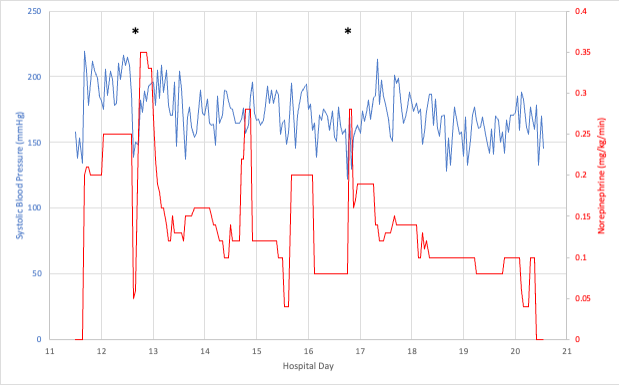

Figure 3: Graph depicts norepinephrine dose and systolic blood pressure measurements over course of induced hypertension. Stars indicate periods where patient symptoms recurred as blood pressure goal decreased.

Discussion

No clinical trial evidence is available to guide the treatment of PCNSV, as many cases are probable but not definitive, and recommendations focus consistently on immunotherapy and overlook acute reversal of cerebral hypoperfusion [4,5,8]. The use of induced hypertension has not previously been reported in the management of PCNSV. In comparison, the use of hemodynamic augmentation is common, albeit controversial, for vasospasm after non-traumatic subarachnoid hemorrhage [9]. As severe multifocal stenosis leading to cerebral ischemia are common to both conditions, it seemed reasonable to trial a higher blood pressure. In this instance, the patient showed clear improvement with vasopressors and declined twice as vasopressors were weaned over 10 days. Our report is the first to support the safety and feasibility of induced hypertension to combat cerebral hypoperfusion in PCNSV.

Conclusion

In PCNSV, induced hypertension may be beneficial to prevent permanent ischemia pending the effect of appropriate immunomodulation.

Disclosure

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Institutional Review Board Statement: IRB approval is not required for case reports by Louisiana State University Health Sciences Center New Orleans. Ethical approval was waived by the local Ethics Committee of Louisiana State University Health Sciences Center New Orleans in view of the retrospective nature of the study and all the procedures being performed were part of the routine care.

Informed Consent: The participant has consented to the submission of the case report to the journal.

Data Availability Statement: Data supporting the study can be provided by request to the corresponding author's email.

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Conflicts of Interest: None

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