A Striking Angiographic Presentation of Primary CNS Vasculitis: A Case Report

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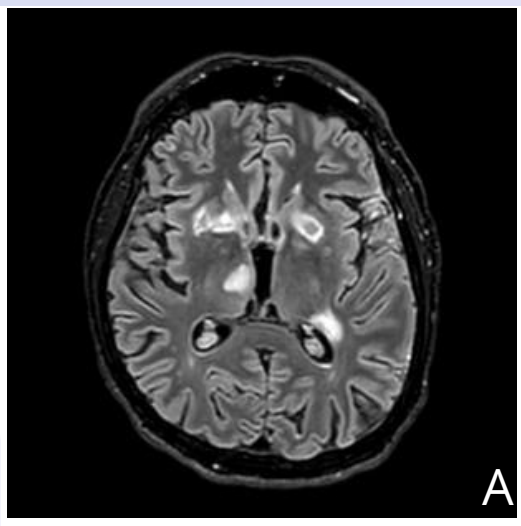
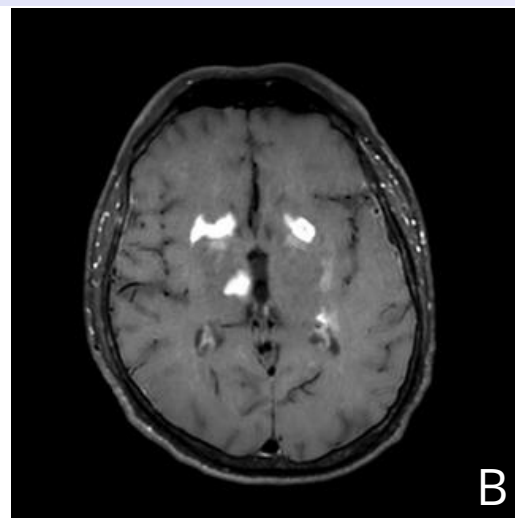
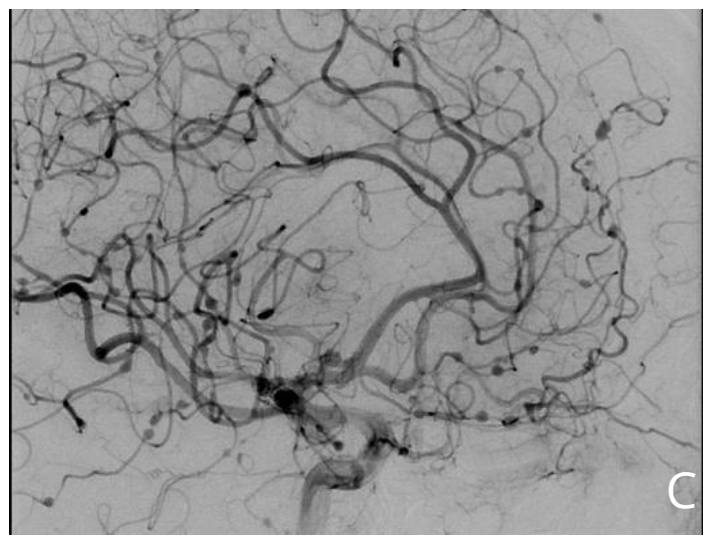
Objectives: Primary angiitis of the central nervous system (PACNS) is a rare, potentially life-threatening vasculitis affecting small and medium-sized cerebral vessels, often presenting with nonspecific neurological symptoms[1] . Early recognition and immunosuppressive treatment are essential to improve prognosis and prevent irreversible damage [3].

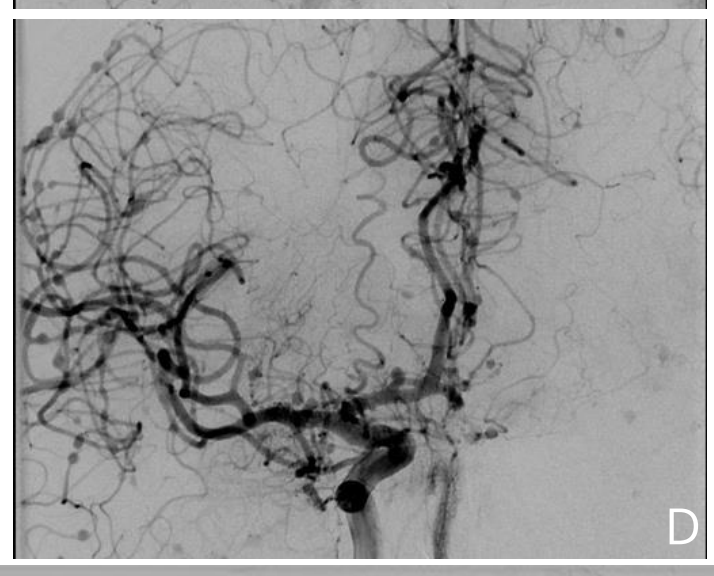
Materials: We evaluated clinical features, neuroimaging, CSF profile, serological and infectious workup, and neuropsychological findings in a middle-aged male admitted to our Neurology department.

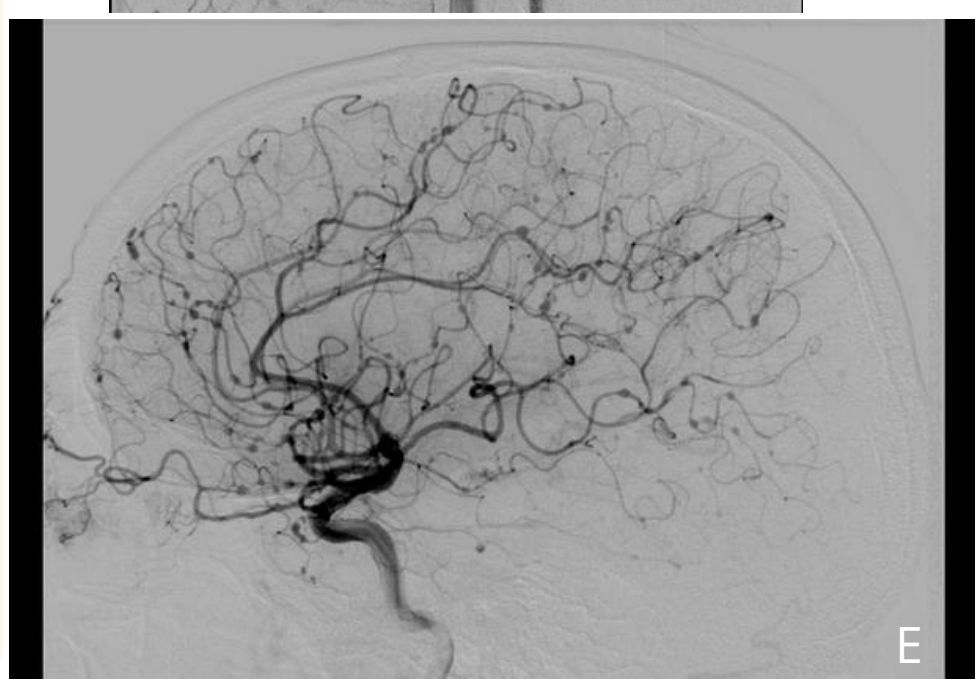
METHOD: A 51-year-old man with no relevant medical history was admitted in February 2025 for acute confusion, lethargy, and behavioral changes. Initial symptoms included fever (up to 40 °C), right otalgia, and facial swelling, initially treated with antibiotics. He subsequently developed progressive disorientation and somnolence. **Brain CT and CTA** showed no hemorrhage but revealed multiple small aneurysms in distal intracranial arteries, raising suspicion of vasculitis.
Neurological exam revealed drowsiness, global disorientation, left facial asymmetry, and mild left hemiparesis. **Lumbar puncture** showed mild hyperproteinorrachia (96 mg/dL), lymphocytic pleocytosis (50 cells/µL), and elevated IgG index (0.75), with negative microbiological and autoimmune panels. EEG showed an asymmetric background with right-hemispheric slowing.











RESULTS: **MRI** revealed multiple acute–subacute ischemic lesions in cortical and deep regions (Images A-B), fusiform aneurysms, and concentric vessel wall enhancement.
Digital subtraction angiography confirmed a "string of beads" appearance and multifocal fusiform dilatations (Images C-D-E), consistent with PACNS. High-dose IV methylprednisolone (1 g/day for 5 days) was started, followed by tapering. A complete rheumatologic workup confirmed PACNS (ESO criteria).
Follow-up MRI with black-blood sequences showed evolving ischemic lesions and persistent wall enhancement. In absence of systemic vasculitis or infection, monthly IV cyclophosphamide was initiated.
Neuropsychological testing at discharge showed mild cognitive slowing and executive dysfunction. The patient achieved full motor recovery.

DISCUSSION: PACNS poses a diagnostic challenge due to its rarity and protean clinical manifestations[1]. This case highlights the diagnostic value of CSF analysis, high-resolution imaging, and angiography. Multifocal ischemic lesions, wall enhancement, and typical angiographic features are key findings. Prompt immunosuppression can halt progression and enable recovery [3].

CONCLUSION: PACNS should be considered in cases of subacute encephalopathy with multifocal ischemic lesions and no systemic involvement. Timely diagnosis and multidisciplinary management are crucial. Further studies are needed to refine diagnostic criteria and treatment protocols.

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