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Revealing Primary CNS Vasculitis in the Shadow of Tuberculosis

Case Report

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Abstract

Introduction: Primary angiitis of the central nervous system (PACNS) is a rare and often under- recognized form of vasculitis confined to the CNS. It typically presents with non- specific neurological symptoms, making it difficult to distinguish from infections, malignancies, and demyelinating disorders, especially in regions endemic for tuberculosis.

Aim: To highlight the diagnostic challenges of PACNS in young individuals and emphasize the importance of a thorough differential workup in cases presenting with recurrent or treatment-resistant neurological symptoms.

Case: A 24-year-old previously healthy male presented with chronic holocranial headache and episodic neurological symptoms including slurred speech and altered sensorium. He was initially diagnosed and treated as a case of tubercular meningitis. However, recurrence of symptoms despite appropriate therapy prompted re-evaluation. MRI findings showed multiple micro-bleeds and perivascular enhancement with MR angiography showing "skipped" or "segmental" pattern of vessel involvement seen — a classical radiological hallmark of PACNS. CSF analysis and serologic workup ruled out infectious and systemic autoimmune etiologies. Based on imaging and exclusion of other causes, a diagnosis of PACNS was made.

Results: The patient was treated with intravenous methylprednisolone followed by oral steroids, low-dose aspirin, and rituximab. He showed significant clinical improvement with no recurrence on follow-up. The case underscores the value of revisiting diagnoses when initial treatment fails, especially in atypical presentations.

Keywords: PACNS, Young Adult; Refractory Headache; Microbleeds; Rituximab; Tubercular meningitis mimics

Introduction

Primary angiitis of the central nervous system (PACNS) is an uncommon and often elusive form of vasculitis, limited to the brain and spinal cord without systemic involvement. First described in the 1950s, PACNS remains a diagnostic challenge due to its rarity, variable clinical presentation, and absence of pathognomonic laboratory findings. The condition can mimic a wide range of neurological disorders, including infections (such as tubercular meningitis), demyelinating diseases, malignancies, and RCVS. PACNS typically presents with sub-acute headache, cognitive disturbances, focal neurological deficits, or seizures. Due to its segmental involvement and lack of systemic markers, the diagnosis is primarily one of exclusion and often delayed.

In India, tuberculosis remains a common CNS pathology, further complicating the diagnosis of PACNS. We present a case of a young male initially treated as tubercular meningitis, who was later diagnosed

with PACNS after relapse of symptoms and detailed re-evaluation. This case highlights the importance of considering PACNS in the differential diagnosis of chronic or relapsing neurological symptoms, particularly in TB-endemic regions.

Case Presentation

A 24-year-old male shopkeeper with no prior comorbidities or addictions presented in July 2023 with persistent holocranial headache of throbbing nature. The headache, initially mild, gradually intensified over weeks. It was associated with blurring of vision but not accompanied by photophobia, phonophobia, vomiting, or diplopia.

He was evaluated at a local clinic, and based on cranial MRI and cerebrospinal fluid (CSF) findings, a clinical diagnosis of tubercular meningitis was made. He was initiated on anti- tubercular therapy (ATT) per DOTS guidelines along with oral dexamethasone. The patient showed initial improvement, with partial resolution of headache symptoms. Repeat MRI with SWI sequencing was done which demonstrated multiple blooming foci in pons, cerebellar vermis and inferior cerebellar cortex. He was subsequently labelled as TB-Reactivation and ATT was continued.

However, in January 2025, he began experiencing episodes of slurred speech, transient altered sensorium, and decreased verbal output with excessive drowsiness. These episodes were spontaneous and self-limiting. Levetiracetam 500 mg twice daily was started empirically.

Despite initial improvement, symptoms recurred on tapering steroids. This prompted further neurological evaluation.

The patient presented to our institution with the above complaints and admitted for further workup.

Examination

On admission, the patient was alert, conscious, and oriented to time, place, and person. Vitals were stable (BP 120/60 mmHg, HR 70 bpm, RR 22/min, SpO_2 98%). There were no signs of systemic involvement—no pallor, icterus, cyanosis, or edema.

- **Neurological examination revealed:**

  Higher mental functions: Intact Cranial nerves: Normal

  Motor system: Normal tone and full power (5/5) in all limbs

  Deep tendon reflexes: 2+

  Plantar responses: Bilateral flexor

  Sensory examination: Within normal limits

- **Investigations**

  Complete Blood Count: Within normal limits

  ESR: Elevated at 70 mm/hr

  Liver and Renal Function Tests: Normal C3 and C4 Levels - within normal limits

- **CSF Analysis (repeat):**

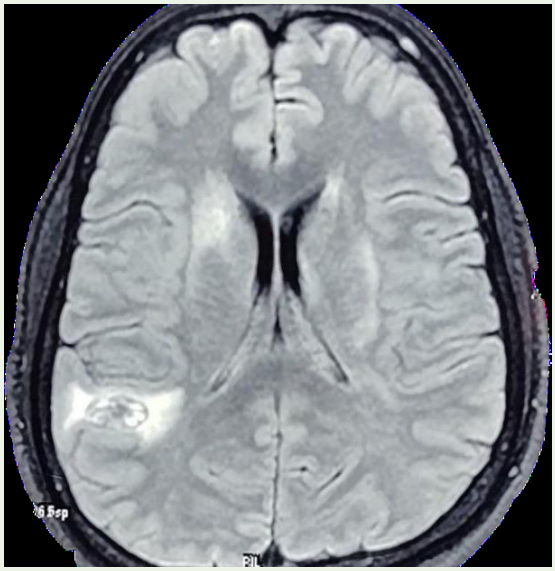

Figure 1: Well defined area of T2/FLAIR of heterogeneous hyper-intensity with surrounding peri-lesional edema noted in right temporo parietal lobe.

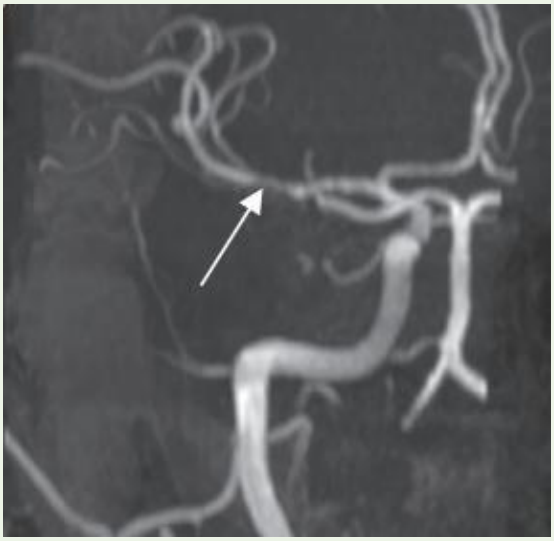

Figure 2: MR - Angio showing, involvement of blood vessels in a "skipped" or "segmental" pattern

Cells: 38 (lymphocytic predominance) Protein: 106 mg/dL

Glucose: 54 mg/dL (RBS: 102 mg/dL) CBNAAT: Not detected

AFB stain: Negative Cryptococcal antigen: Negative

CSF for Malignant cells: Negative

- **Autoimmune Panel:**

  ANA: Negative

  p-ANCA and c-ANCA: Negative Anti-AQP4 / Anti MOG: Negative

  Rheumatoid Factor and Anti-CCP: Negative

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MRI Brain (sequential) with MR Angio and Whole Spine Screening:

Intra cerebral hematoma in right parieto occipital lobe with peri-lesional edema

Multiple micro-bleeds and punctate microhemorrhages dispersed in bilateral brain parenchyma

Confluent T2/FLAIR hyperintensities in bilateral deep cerebral white matter.

Post-contrast imaging: Perivascular enhancement along vessels in bilateral cerebral hemispheres

MR angiography: "Skipped" or "segmental" pattern of vessel involvement seen — a classical radiological hallmark of PACNS. This refers to alternating areas of stenosis and normal-caliber segments across multiple intracranial vessels, reflecting multi focal vasculitic inflammation.

MRI Spine Screening - With in normal limits with no demyelinating lesions

After comprehensive evaluation, infectious etiologies such as TB Meningitis, Neuro-syphilis and HIV related Vasculopathy were ruled out with negative CSF CBNAAT, negative AFB staining, absence of basal meningeal enhancement, negative VDRL serologies and negative HIV serologies respectively.

Auto-immune mimics such as SLE, ANCA vasculitis, Rheumatoid vasculitis and Bechet's were ruled out as ANA, ANCA, RA, anti CCP and C3, C4 levels were not suggestive along with absence of any systemic sign/symptoms such as absence of any rash, hematuria, hemoptysis, oral or genital ulcers.

Other Demyelinating diseases such as MS, NMO and MOG were ruled out with the absence of any demyelinating lesions on MRI and absence of Anti Aquaporin and MOG antibodies.

Based on imaging and exclusion of other differentials—including CNS infections, Autoimmune /Systemic vasculitis, and Demyelinating disorders—a diagnosis of Primary Angiitis of the Central Nervous System (PACNS) was established.

Treatment and Follow-up

The patient was started on:

* Intravenous methylprednisolone pulse therapy (1 g/day for 3 days)
* Maintenance oral corticosteroids (Prednisolone)
* Aspirin 75 mg once daily
* Rituximab-based immunosuppressive therapy

The patient showed marked clinical improvement following immunosuppressive therapy. At follow-up visits, there were no further episodes of headache or altered sensorium. Neurological examination remained normal, and he resumed daily activities without limitations. Continued follow-up is planned to monitor for disease recurrence or complications related to immunosuppressive therapy.

Discussion

Primary angiitis of the central nervous system (PACNS) is an elusive and often misdiagnosed condition, primarily due to its non-specific presentation and its overlap with more prevalent neurological disorders, especially in tuberculosis (TB)-endemic regions. Our case underscores the diagnostic complexity of PACNS and the critical importance of maintaining a high index of suspicion when patients fail to respond to standard therapies.

The initial misdiagnosis of tuberculous meningitis in our patient reflects a common clinical pitfall in regions like India, where TB remains a major public health concern. In this case, prolonged anti-tubercular therapy (ATT) failed to produce sustained improvement, and symptom relapse upon steroid tapering prompted reconsideration of the diagnosis.

The turning point in diagnosis was the detailed review of sequential MRI findings, which revealed microhemorrhages in the cerebellum and brainstem, along with perivascular enhancement and MR angiography revealing skipped and segmental pattern of vessel involvement — features more characteristic of PACNS than TB. The cerebrospinal fluid (CSF) analysis, while showing elevated protein and lymphocytic pleocytosis, lacked evidence of TB, malignancy, or systemic autoimmune disease. Negative CSF CBNAAT and ADA levels further strengthened the case against a tuberculous etiology.

Histopathological confirmation through brain biopsy remains the gold standard for PACNS diagnosis but is often impractical due to the invasiveness and location of lesions. In such cases, a diagnosis of exclusion based on clinical judgment, imaging, and laboratory data becomes essential. This aligns with current diagnostic frameworks that prioritize non-invasive tools when biopsy is not feasible.

Therapeutically, the patient's significant improvement with immunosuppressive treatment — including intravenous methylprednisolone and rituximab — reaffirms the autoimmune nature of PACNS and the need for timely initiation of appropriate therapy.

Conclusion

This case exemplifies the diagnostic challenge posed by Primary CNS Vasculitis (PACNS), particularly in TB-endemic regions where infectious etiologies are often the first consideration. The patient's prolonged misdiagnosis as tuberculous meningitis, despite atypical features and poor therapeutic response, underscores the need for heightened clinical vigilance.

Early recognition of PACNS requires a combination of detailed neuroimaging, exclusion of mimics, and a multidisciplinary approach. This case also highlights the value of immunosuppressive therapy — especially the role of rituximab — in achieving favorable outcomes. Clinicians must consider PACNS in the differential diagnosis of young adults with chronic relapsing neurological symptoms unresponsive to conventional treatments.

Timely diagnosis and initiation of appropriate therapy are critical to preventing irreversible neurological damage and improving patient prognosis in this rare but treatable condition.

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Authors' Contribution

All authors contributed equally to patient care, data collection, literature review, and manuscript preparation. Dr. Pranita, Dr. Wasnik P and Dr. Shukriya S led the case diagnosis and treatment strategy along with supervised clinicaal decisions. All authors reviewed and approved the final manuscript.

Ethical Compliance: All procedures performed in this case were in accordance with the ethical standards of institutional and/or national research committee and with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards.

Conflict of Interest declaration: The authors declare that they have no conflict affiliations with or any organization or entity with any financial interest in the subject matter or materials discussed in this manuscript. The authors have no conflicts of interest to declare.

Consent: Written and Informed consent of the patient and all authors were taken for publication of this case.

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