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A CASE REPORT OF ACUTE LACUNAR STROKE IN A PATIENT WITH PRIMARY CNS ANGIITIS

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ABSTRACT

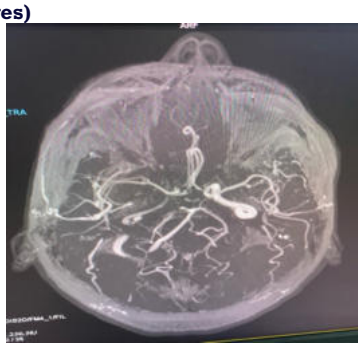
Primary CNS angiitis is a rare and challenging condition characterized by inflammation of blood vessels in the central nervous system. It has a male preponderance with a median onset of 45 years.1 Patients usually present with combination of symptoms like headache, seizure and focal neurological deficit associated with destructive changes, occlusion and infarction.2 The term primary angiitis of central nervous system (PACNS) unifies a group of clinico-pathological presentation of vasculitis affecting the vessels of brain and spinal cord without any overt systemic vasculitis of underlying potential cause.3 In 'primary' or 'isolated' vasculitis or angiitis of the CNS, there is little or no overt generalized inflammation while in 'secondary angiitis' the CNS becomes involved in a systemic vasculitic illness like Wegener's granulomatosis or microscopic polyarteritis.4 More than 25 years after the introduction of the first diagnostic criteria for PACNS by Calabrese and Mallek, it's diagnosis still remains challenging as there is a long time lag between the symptom onset and eventual diagnosis.5 Even though a plausible diagnosis of PACNS may be made on MRI corelating with clinical findings and cerebral DSA, meningo-cortical or spinal cord biopsy remains the reference standard to certify the diagnosis but is relatively inaccessible and potentially hazardous.6 All forms of cerebral vasculitis are relatively rare but all are serious and potentially life-threatening.

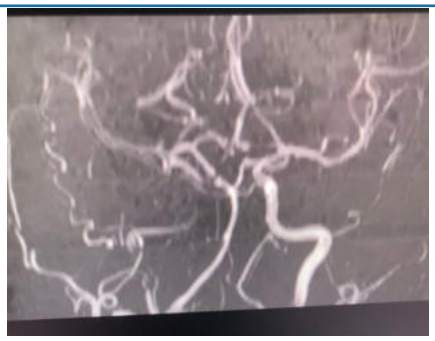
CASE REPORT

A 42 year old male patient presented to the Emergency Department with complaint of headache after five minutes of which he developed giddiness both sudden in onset and non-progressive in nature. Few minutes later, he developed slurring of speech followed by left upper limb weakness where he was unable to lift even a paper for a few minutes without any associated weakness in lower limbs post which it resolved and he could drive his car to home. After 15 minutes of which, he developed tingling and numbness in left upper limb. He occasionally drank alcohol but had an otherwise healthy lifestyle. Family and past history was unremarkable.

On presentation, his vitals were within normal limits. His physical examination revealed he was conscious, alert and oriented to time, place and person. His respiratory, cardiac and abdominal examination was within normal limits. His CNS examination revealed power in left upper limb in hand grip, wrist and elbow was 4/5 which improved remarkably after physiotherapy. On evaluation, his all routine investigations like complete blood count, liver function tests and kidney function tests were well within normal limits. His MRI Brain was suggestive of and MR Angiogram revealed which was suggestive of CNS vasculitis so antineutrophilic cytoplasmic antibodies c-ANCA and p-ANCA were done which were normal, ruling out the possibilities of necrotizing vasculitis like Wegener's granulomatosis, polyarteritis nodusa, churgstrauss syndrome etc.

(MRI Images)





The patient was then admitted to the neurology ward where his initial treatment was started for stroke with and was further treated with. Over time, the patient's power in left upper limb improved to 5/5, his headache resolved and was discharged home symptom free.

DISCUSSION

The diagnostic criteria for PCNSV were initially proposed by Calabrese and Mallek (1988) and included the following: (a) the presence of unexplained neurological deficit after thorough clinical and laboratory evaluation; (b) documentation by cerebral angiography and/or biopsy revealing arteritis in the CNS; and (c) and the absence of systemic vasculitis or other secondary condition associated with the angiographic or pathological features.[7]

Brain biopsy remains the gold standard in the diagnosis of PCNSV, but patchy or inaccessible lesion involvement may result in false negatives. Angiography is considered the gold standard in the imaging diagnosis of PCNSV. Currently, a diagnostic sensitivity of 90-100% is recognized for brain MRA, and its value in the diagnosis of angiitis is increasing.[6,8]

Our patient revealed a focal neurological deficit, and the initial imaging findings were suggestive of an ischemic stroke. Vascular occlusive causes of stroke were ruled out in the patient with no other identified vascular risk factors, except for stage 1 hypertension which was diagnosed and treated during hospitalization.

The presence of multiple stenoses across different vascular territories observed on brain imaging, along with the findings from transcranial Doppler ultrasound, strongly indicated the presence of vasculitis(narrowing of vessels on MRA)

Secondary causes of CNS vasculitis such as infections, systemic vasculitis, autoimmune diseases, and/or lymphoproliferative diseases were excluded. The combination of unexplained recurrent neurological deficits, angiographic features indicative of CNS angiitis, and the absence of systemic vasculitis are established diagnostic criteria for PCNSV.

Because a brain biopsy was not performed, histological information regarding an inflammatory process associated with vasculitis could not be obtained. Therefore, the possibility of RCVS(Reversible cerebral vasoconstriction syndrome) cannot be completely ruled out. This condition is equally rare and shares similar clinical and imaging features with PCNSV.

In RCVS, the primary manifestation is a sudden and intense thunderclap-like headache.[9] The clinical presentation of RCVS can vary widely in terms of severity and may be accompanied by hypertension and focal neurological deficits. The etiopathogenesis of RCVS in relation to ischemic stroke remains unclear. Histopathological studies in patients with RCVS document minimal vascular changes, pointing to functional vasoconstriction rather than a vasculitis phenomenon characteristic of PCNSV, highlighting the increasing role of brain MRA as a non-invasive diagnostic option.

The diagnosis of PCNSV holds importance due to its unique therapeutic approach, which differs from the management of other common causes of ischemic stroke. This approach typically involves the administration of systemic glucocorticoids, with a lower risk of symptom recurrence if used in combination with cyclophosphamide. While in PCNSV there is a favourable response to corticosteroid therapy, it should be avoided in RCVS because its use is considered an independent predictor of a worse prognosis.

Our patient had no other known vascular risk factors and no signs of atheromatosis in the cervical vessels. In addition to neurological deficits, she presented with symptoms of a mild-to-moderate and self-limiting holocranial headache.[9] Imaging studies revealed the presence of multiple intracranial stenoses, which were suggestive of vasculitis. Finally, the patient exhibited a positive response to corticosteroid therapy, with a favorable clinical outcome. Therefore, PCNSV was considered the underlying cause of the ischemic stroke, rather than RVCS.[10]

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