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NEW ONSET STATUS EPILEPTICUS AND MULTIFOCAL HAEMORRHAGE IN A YOUNG ADULT, A RARE CLINICAL PRESENTATION OF CNS VASCULITIS

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ABSTRACT

Primary central nervous system vasculitis (PCNSV) is a rare inflammatory disorder affecting the blood vessels of the brain and spinal cord, without systemic involvement or underlying connective tissue disease. It usually presents with a wide range of nonspecific symptoms in the form of focal neurological deficits, new onset seizures, severe headaches, and cognitive impairment. Diagnosis is usually made when unexplained neurological deficits with either an angiogram or CNS biopsy shows features of vasculitis with other causes ruled out. Few cases are being reported every year thus making the diagnosis of PCNSV difficult in emergency department. The patient discussed here is a 29-year-old male who presents as status epilepticus to our emergency department. MRI brain reveals multiple subacute intraparenchymal haemorrhage with punctate micro haemorrhages. CSF analysis shows pleocytosis with mildly elevated protein levels. A diagnosis of primary CNS vasculitis was made and patient rapidly improved after receiving intravenous

Methylprednisolone for 7 days.

KEYWORDS: Primary CNS vasculitis (PCNSV), Status Epilepticus, Methylprednisolone, Intraparenchymal haemorrhage.

INTRODUCTION

Primary central nervous system vasculitis (PCNSV) is an uncommon inflammatory condition selectively affecting blood vessels of the central nervous system. Its presentation can be varied and diagnosis remains difficult. Untreated, PCNSV can progress with catastrophic effects due to multiple infarcts and bleeds. Early, diagnosis and treatment improve outcomes.[1] It has a male preponderance with a median onset of 50 years. Patients usually present with a combination of headaches, seizures and focal neurological deficits, and there is a long-time lag between symptom onset and eventual diagnosis. The diagnosis is made by the Calabrese and Mallek criteria, where an unexplained acquired neurological deficit with either an angiogram or CNS biopsy shows features of vasculitis, with other causes ruled out and requires a high index of suspicion.[2] Imaging and laboratory data can aid diagnosis using brain MRI, angiography, and CSF examination. Brain MRI abnormalities are reported in up to 96% of patients. Angiography is another leading tool to help in the diagnosis with an estimated sensitivity of 50 to 90%. Typical MRI findings show multifocal areas of signal intensity in both the white and gray matter, the presence of gadolinium-enhancing lesions, and evidence of both acute and chronic strokes. Angiography findings often show segmental narrowing, beading, or dilation of vessels, although sensitivity can vary. Typical CSF findings include mild lymphocytic pleocytosis, elevated protein levels, and occasionally oligoclonal bands, indicative of an inflammatory process. Brain tissue biopsy demonstrating angiitis remains the gold standard.[3]

CASE PRESENTATION

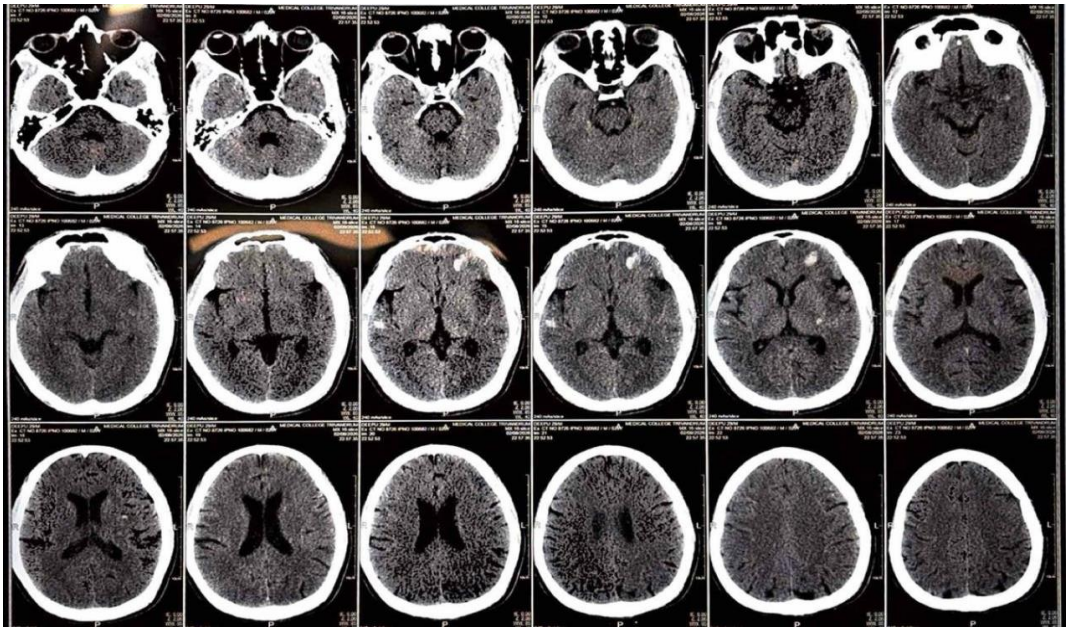
A 29-year-old male with no known comorbidities presents to the emergency room with episodes of three generalised tonic clonic seizures, up rolling of eyeball and urinary incontinence without regaining consciousness in between the episodes. There is no history of headache, fever, vomiting, giddiness or trauma. There is no history of similar complaints in the past or family history of any autoimmune diseases. On examination pulse rate of 100 beats per minute and Spo2 of 88 % in room air with BP of 130/70 mm Hg was observed. There was no pallor, cyanosis, icterus, edema or peripheral lymphadenopathy. Chest examination reveals bilateral equal air entry with no basal crepitations. CNS examination

reveals bilateral pupil sluggishly reactive to light. The cranial nerve, sensory and motor system examination were normal. There were no signs of cerebellar dysfunction or meningitis. Tone was normal bilaterally with bilateral flexor plantar response. In view of poor GCS (E1V1M3) and threatened airway, patient was intubated. The seizures were refractory to the maximum loading dose of lorazepam and levetiracetam but was controlled with midazolam infusion.

Investigations

CBC shows leucocytosis with neutrophilic response. Random blood sugar, renal function and liver function parameters were within normal limits. Cerebrospinal fluid (CSF) analysis revealed mild inflammatory changes, including pleocytosis and mildly elevated protein (49 mg/dL), with negative infective serology panel ruling out active infection. EEG shows mild degree of generalised electro physiological disturbance in the form of delta range slowing with no epileptiform abnormalities. Autoimmune profile was also negative.

An initial non-contrast head CT revealed multiple intraparenchymal hemorrhages in the frontal and temporal lobes. Subsequent CT angiogram and venogram showed normal arterial and venous circulation without evidence of thrombosis or occlusion. MRI of the brain demonstrated multiple subacute Intraparenchymal haemorrhage in the bilateral temporal and left frontal lobes with extensive bilateral asymmetric white matter T2/FLAIR hyperintensities involving frontotemporal regions with multiple punctate microhemorrhages.



The NCCT head is shown above. Brain biopsy was deferred by the patient.

A diagnosis of CNS vasculitis was made, and the patient received IV Methylprednisolone 1g/day for 7 days and showed significant improvement after that. Further hospital stay of the patient was uneventful with full neurological recovery and was discharged with neurology follow up.

DISCUSSION

Primary angiitis of the central nervous system (PACNS) is a rare variant of vasculitis that is restricted to CNS which has to be considered as one of the differential diagnoses in young adults presenting with status epilepticus. Due to varying clinical presentation and radiographic characteristics diagnosis of PACNS remains as a diagnostic challenge. The etiology and pathogenesis of PACNS remains unclear. As in all vasculitis, the pathogenesis is thought to be immunological in nature. Crowe has proposed four different mechanisms of tissue injury that can occur in vasculitis: deposition of immune complexes, direct antibody mediated damage, delayed hypersensitivity and cytotoxic T lymphocytes mediated damage.[4] The criteria for diagnosis have been laid down by Calabrese and Mallek.[5] But due to its heterogeneous presentation clinicians still find it difficult to diagnose in day-to-day practice. It is a disease that shows slight preponderance in men, with a male to female ratio of 4 to 3. Most patients in the literature had a diverse presentation, but headache and cognitive decline were the most common reported symptoms at presentation. Although less common, seizures were reported in the range of 7–29% as discussed in our case.[6] Vascular events are the hallmark of the disease process. Clinical presentation is often due to recurrent small strokes. About 30-50% patients with PCNSV present with acute stroke. PCNSV is associated with increased intracranial hemorrhage and thrombolysis is contraindicated.[1]

PCNSV should be added to the check list of contraindications for thrombolysis and emergency care physicians involved in acute stroke management should be aware of the condition.[1] Cerebrospinal fluid findings play a crucial role in the diagnosis of PACNS. It is abnormal in 80–90% in whom PACNS was pathologically confirmed.[6] In our case the patient's CSF analysis was abnormal and showed pleocytosis with an elevated protein. CSF examination is extremely useful in the exclusion of possible causations of the patient's status epilepticus. The CSF examination in our case was negative for any infectious, and autoimmune antibodies suggesting an underlying alternative pathology. Certain MRI features should raise suspicion of PCNSV. Multifocal ischemic and hemorrhagic lesions of different

ages involving gray and white matter of different Vascular territories, subarachnoid hemorrhage (SAH) noticeable as sulcal FLAIR signal changes, patchy areas of enhancement, leptomeningeal, dural and vessel wall enhancement were some of the common ones observed.[1] MRI of the brain in our case demonstrated multiple subacute IPH in the bilateral temporal and left frontal lobes, alongside extensive asymmetric white matter T2/FLAIR hyperintensities, punctate microhemorrhages, and perivascular space enhancement. Brain biopsy still remains as gold standard for diagnosis of PCNSV.[7] The diagnostic yield and safety of brain biopsy have been well studied.[8] Steroids with cyclophosphamide or azathioprine is the recommended treatment regimen. The combination of steroids with rituximab, methotrexate, mycophenolate has also been shown to be effective. The treatment of PCNSV is always guided by expert opinion as relapse on treatment has been known to occur.[9,10]

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