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Case Report

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CNS VASCULITIS PRESENTING AS STATUS EPILEPTICUS

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ABSTRACT

Primary central nervous system vasculitis (PCNSV) is a rare inflammatory arteriopathy confined to the brain, spinal cord, and leptomeninges. Because of its nonspecific presentation and difficulties in making a positive diagnosis, empiric treatment is often instituted. Diagnostic criteria include newly acquired neurological deficits, unexplained by another central nervous system (CNS) or systemic process, in the presence of a highly suggestive angiogram and/or biopsy. There are few cases each year making further characterization and correct diagnosis difficult. The patient discussed here is a 65 year old male who was admitted as a case of status epilepticus in our casualty. Patient was found seizing on the roadside by some passers by and was brought to our hospital in unconscious state. MRI brain revealed large infarcts in the distribution of right MCA and ACA territory. CSF was cellular with lymphocytic pleocytosis and elevation in proteins with normal sugars. A diagnosis was primary CNS vasculitis was made and patient received IV Methylprednisolone for 5 days and improved significantly.

KEYWORDS: Primary CNS vasculitis (PCNSV), Methylprednisolone, immunosuppression.

INTRODUCTION

Primary central nervous system vasculitis (PCNSV) is a rare inflammatory arteriopathy confined to the brain, spinal cord, and leptomeninges. True PCNSV is devastating. It is typically insidious in onset and progressive, presenting around the fourth decade of life. Classically, patients report progressive headache and encephalopathy. They have multiple radiographic lesions in varying vascular distributions on neuroimaging that can be both ischemic and haemorrhagic in nature. The most common symptoms of vasculitis are relatively nonspecific, the average time to diagnosis can be several months. Given the presence of ischemia and/or hemorrhage, CNS vasculitis is often included on the differential diagnosis of stroke. A major diagnostic challenge is the similarity between CNS vasculitis and other more common disease mimics. Intracranial atherosclerosis and reversible cerebral vasoconstriction syndrome (RCVS) are far more common disorders and often misdiagnosed, as we illustrate below. Other mimics include CNS lymphoma, neurosarcoidosis, multiple sclerosis, fibromuscular dysplasia, hypercoaguable states (ie, antiphospholipid antibody syndrome), amyloid angiopathy, and malignancy. CNS infections and changes secondary to drug exposure and radiation can also have similar clinical presentations and imaging findings. The frequency of their occurrence is significantly higher than CNS vasculitis.

CASE PRESENTATION

A 65 year old male was brought to casualty with history of generalised tonic clonic seizures lasting for about 30 minutes followed by prolonged post ictal phase. Patient was received in the casualty in post-ictal state which lasted for about 10 hours.

On examination patient was deeply comatosed, decerebrate postured. Pulse of 88 beats per minute and all the peripheral pulses were felt, BP of 150/90 mmHg, an SPO2 of 85%. There was no pallor, icterus or edema, but patient was cyanosed. There was no palpable lymphadenopathy. Chest examination revealed bilateral coarse crepitations all over the chest and there was shallow rapid breathing. CVS and abdominal examination was grossly normal. CNS exam revealed decebrate posturing and pupils were bilaterally pinpoint. There was no facial asymmetry. Motor system examination revealed that bulk was normal in all the four limbs. Tone was slightly increased on right side. Power could not be assessed. DTR were normally present. Left sided plantar showed extensor response while right showed normal flexor response. There were no cerebellar or meningeal signs.

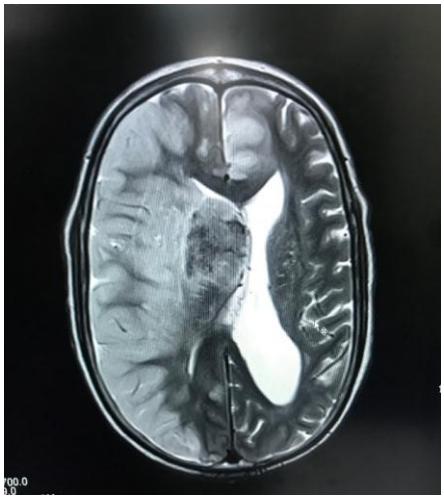
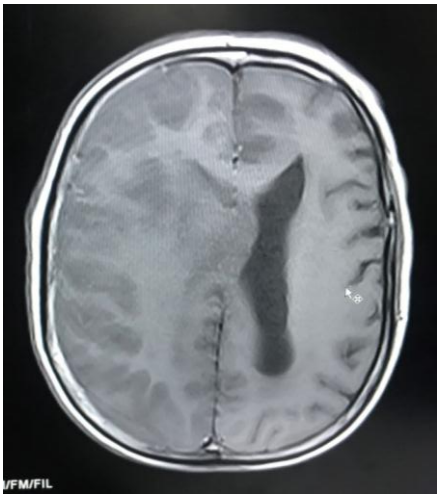
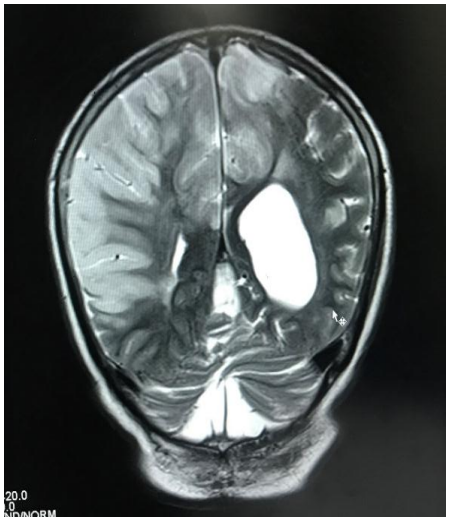
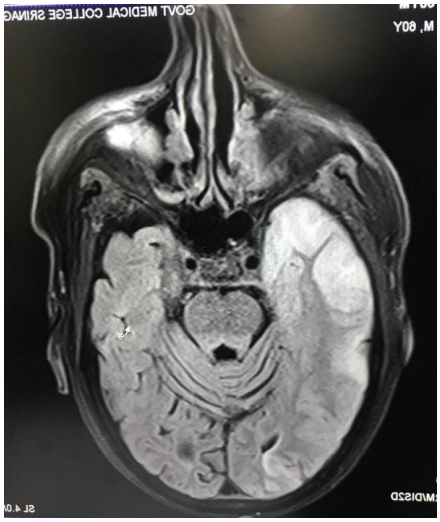
Investigations

- CBC with differential – leucocytosis with neutrophilic response
- KFT – normal, LFT- normal, ESR of 68mm in 1st hour

- Lipid Profile – normal
- EEG – suggestive of global encephalopathy
- USG abdomen/pelvis- normal
- NCCT head- mild corticocentral atrophy with no evidence of any infarct or haemorrhage (likely early CT).

- CSF analysis-

TLC	DLC	Protein	Sugar	Staining	MTB PCR	Autoimmune profile
11	N 10 L 90	78mg/dL	60mg/dL	Negative	Negative	Negative

Brain biopsy was refused by the patient.

A diagnosis of CNS vasculitis was made on the basis of history, MRI and CSF findings. Patient received IV Methylprednisolone 1g/day for 5 days and showed significant improvement after that. Though patient improved in view of sensorium, patient was still hemiplegic and continued with aphasia. He was put on maintenance dose of steroids and was discharged in hemodynamically stable condition. Patient is on regular neurology follow up from last 2 weeks.

DISCUSSION

Primary angiitis of the central nervous system (PACNS) is frequently considered in the differential diagnosis in patients with cryptogenic neurologic illness or in young subjects with ischemic stroke. The absence of characteristic clinical, laboratory, or radiographic features of this rare disease make the diagnosis very difficult, and has contaminated the literature with unproven cases in which alternative diagnoses are plausible. The etiology and pathogenesis of PACNS are unknown. The fundamental mechanism of all vasculitides is immunologic; Crowe discussed 4 different mechanisms of tissue injury that might apply to the pathogenesis of vasculitis: immune complexes, direct antibody-mediated damage, delayed hypersensitivity, and cytotoxic T lymphocytes.[1] With the limited knowledge we have about PACNS, no strong evidence supports any of these mechanisms in the pathogenesis of this disease, although the granulomatous nature of inflammation suggests a role of cell-mediated immunity.[2] As in other autoimmune disorders, T cells that become sensitized in the course of systemic illness or viral infection probably later contribute to a cellular immune response directed against cross-reacting epitopes in CNS vessels.[3] Other authors propose that, in the setting of altered host defense mechanisms, a virus or other pathogen may lead directly or indirectly to diffuse cerebral vasculitis.[4]

The latter hypothesis is supported by the rare condition in which vasculitis involving mainly the ipsilateral anterior circulation with consequent infarcts occurs days to weeks following varicella-zoster infection of the first division of the trigeminal nerve. The mechanism seems to be retrograde spread of the virus from Gasserian ganglion to the arteries of the anterior circle of Willis.[5] Pathologically confirmed cases of PACNS have been reported in patients with Hodgkin disease, amyloid angiopathy, and graft-versus-host disease. However, available information in these cases does not allow any conclusion about the causal relation between these diseases and PACNS.[2] Regardless of the etiology of PACNS, the main mechanism of neurologic damage in these patients is ischemic. This results from 3 consequences of inflammation in the vascular wall: obstruction of the vessel lumen, increased local coagulation from the effects of proinflammatory cytokines on the endothelial surface, and alteration in vasomotor tone.[6]

Mortality and morbidity of PACNS are hard to determine due to the variability in diagnosis means and treatment among published series. However, treatment with steroids and immunosuppressants has improved the outcomes of the disease, which used to be fatal. In a recent report, 14% of 29 patients with biopsy-proven PACNS died or had severe morbidity (Modified Rankin Scale of 5) at follow-up of 1.14 years.[7]

Men are more commonly affected by PACNS than women; the male-to-female ratio is about 7:3.[4] In most reported cases, patients were in the fourth to sixth decades of life at time of diagnosis. However, patients aged 7 months to 78 years have been described.[8,9,10] Although it is nonspecific, CSF is abnormal in most patients with PACNS. Mild-to-moderate CSF pleocytosis, predominantly lymphocytes, is found in approximately 70-90% of cases.[2,14] The vast majority of patients have elevated CSF protein, with a median protein concentration of 98 mg/dL (range 44-1034 mg/dL) in a recent report.[14] In some patients, oligoclonal bands can be detected in the CSF, reflecting an inflammatory process in the CNS. The glucose is usually normal. Cultures and cytology studies are important to exclude CNS infection and malignancies. In some patients, initial CSF examination results might be normal, but subsequent lumbar punctures show abnormalities as the disease evolves. CBC, antinuclear antibodies (ANA), RF, SSA/SSB, p-ANCA, c-ANCA, cryoglobulin, complement levels, other tests for rheumatic diseases, and syphilis serology usually produce normal results but are helpful to rule out other conditions that can mimic PACNS.ESR is usually normal or mildly elevated. Head CT shows various combinations of nonspecific findings, including cerebral infarcts or hemorrhages with mass effect, hydrocephalus, or white matter hypodensity. CT may also be normal. MRI of the brain is essential in the evaluation of patients with possible PACNS; it helps to exclude other diagnoses such as CNS tumors, demyelinating diseases, multiple small infarcts, and hydrocephalus. Brain biopsy is the only method able to establish a diagnosis of PACNS and to rule out other diseases. Because vessels in leptomeninges are involved in most cases, the leptomeninges should always be sampled. Lesions seen on MRI should be biopsied if accessible; otherwise, the nondominant frontal or temporal lobe can be sampled. Both open and stereotactic biopsies have been used with similar outcomes.

Combined immunosuppressive therapy is the treatment of choice for PACNS. Current recommendation is oral prednisone, 1 mg/kg/d, and cyclophosphamide, 2 mg/kg/d. Treatment consists of 2 phases: Induction of remission and maintenance of remission. Duration of each phase is debatable, but most centers use prednisone and cyclophosphamide for 4-6 months to induce clinical remission, and then taper prednisone off to continue cyclophosphamide for 1 year.

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