CASE REPORT

PRIMARY ANGIITIS OF CENTRAL NERVOUS SYSTEM

Nilesh Kumar[1], Dibya R Behera[2], Ankur N Varshney[2], Arvind Anand[2], NK Singh[3]

[1]Senior Resident, [2]Juniour Resident, [3]Professor, Department of General Medicine, IMS, BHU, Varanasi, India

Correspondence: Dr. Nilesh Kumar, Email: nilesh19arreno@gmail.com

ABSTRACT

Primary angiitis of central nervous system (PACNS) is a rare disorder. We report a 20 year old boy presented with sudden weakness of left side of body with altered sensorium with background of headache for 15 days. MRI and MR angiography revealed feature suggestive of PACNS which was supported by relevant blood investigations. The case was treated with cyclophospamide and methylprednisolone and had shown improvement.

Keywords: Primary Angiitis, Central Nervous System, Altered Sensorium

CASE

A 20 year old male presented with headache for 15 days and sudden weakness in left half of body followed 15 min. by weakness in right side of body associated with altered sensorium. He had no history of head trauma, fever, seizure, joint pain, oral ulceration, rash, digital infarct. There was no past history of similar illness; neither any family member had such illness.

He was admitted in comatose state with pulse rate 78/min & BP 110/ 70 mmHg. On motor system examination, he had normal bulk in all 4 limbs, increased tone and brisk reflexes in all 4 limbs. Plantar reflex was not illicitable. All other systems were within normal limits.

His routine investigations including CRP and ESR were within normal limits.

NCCT head showed multiple ill defined areas of hypodensity in pons, posterior limb of Right internal capsule, bilateral peri-ventricular white matter, predominantly posteriorly which is a sigh of ischemia.

CSF was normal and no oligoclonal band was seen.

MRI Brain showed multiple ischemic infarcts at various stages of evolution seen in medulla, pons left cerebellar hemisphere, vermis, right basal ganglia, left thalamus and peri ventricular white matter in bilateral frontal and parietal lobes- feature suggesting vasculitis with multiple infarcts (Figure-1a,1b,1c).

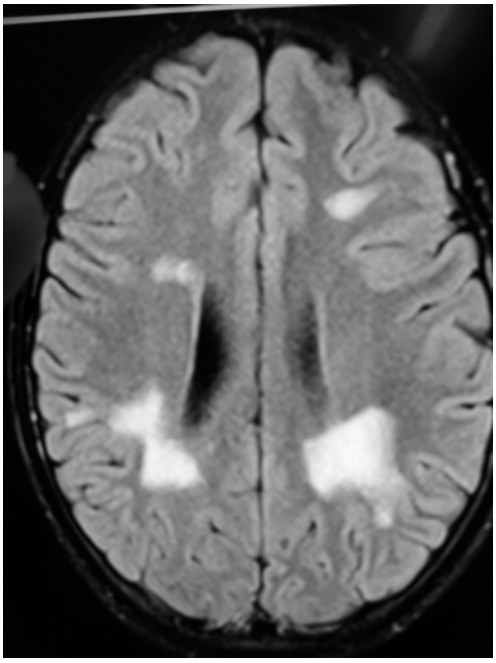

Figure 1a

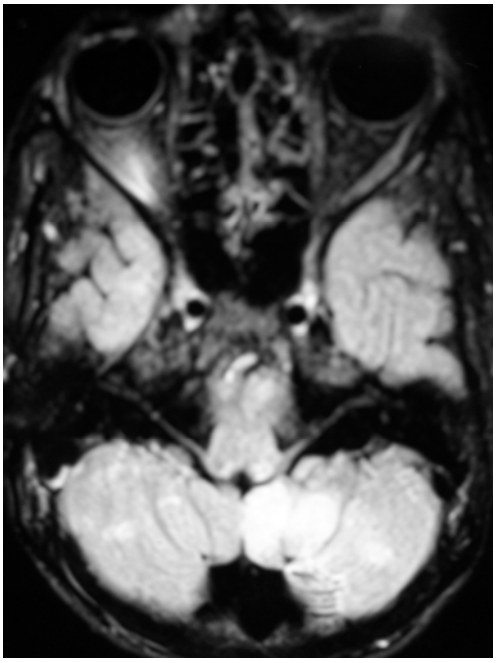

Figure 1b

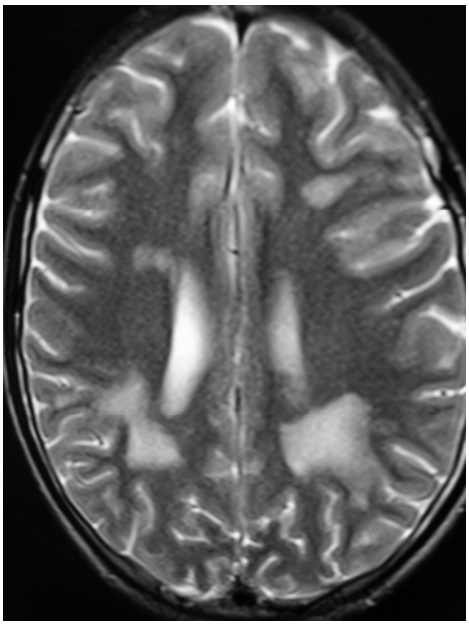

Figure 1c

Fig 1a, 1b & 1c: MRI Brain showed multiple ischemic infarcts at various stages of evolution seen in medulla, pons left cerebellar hemisphere, vermis, right basal ganglia, left thalamus and peri ventricular white matter in bilateral frontal and parietal lobes

MR Angiography of brain showed significant narrowing of distal part of basilar artery (Figure-2). Following investigations were found to be within normal limits-ANA, Anti ds DNA, RA factor, Antiphospolipid antibody screen, coagulation profile, LDH. Multiple blood cultures, HIV, HBsAg, HCV were negative.

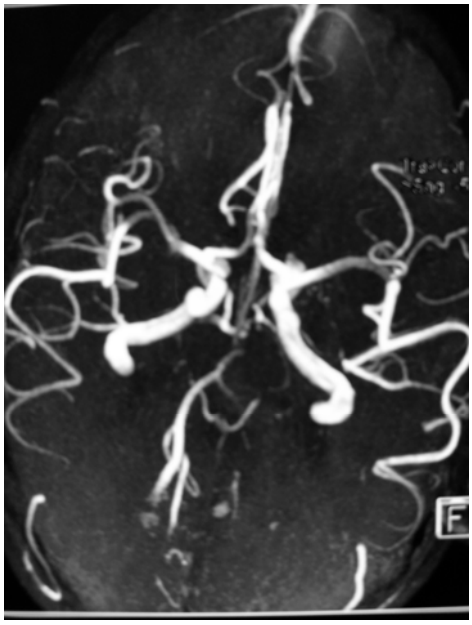

Fig 2: MR Angiography of brain showed significant narrowing of distal part of basilar artery

A provisional diagnosis of primary CNS angiitis was considered. Treatment was initiated with cyclophosphamide 50 mg per oral BD and prednisolone 20 mg per oral BD. After one month, though he had some improvement in form of flickering of fingers, he presented with severe breathlessness for which he was intubated. Treatment plan was reviewed and decided that monthly pulse of inj. Cyclophosphamide 600mg/ m2 BSA with inj. Methylprednisolone 1 gm to be given for six months. Following the first pulse he showed dramatic improvement like - extubated in 8 days, able to hold objects, lifting hand above head, able to swallow foods, able to sense urination and defecation. Now he is in follow up and doing well.

DISCUSSION

Primary angiitis of the central nervous system (PACNS), a rare form of vasculitis restricted to the brain and spinal cord. Diagnostic criteria of primaiy angiitis of CNS were proposed in 1988. These criteria included (i) an unexplained neurologic deficit despite aggressive diagnostic evaluation; (ii) a high probability angiogram for arteritis and/or histopatliologic evidence of arteritis confined to the CNS; and (iii) exclusion of all those disorders capable of mimicking the angiographic findings or associated with vascular inflammation of the CNS[1]. The clinical presentations are highly variable but the triad of headache. organic brain syndrome and multifocal neurodeficit is highly suggestive of the condition. Systemic symptoms are mostly absent in the affected patients as is laboratory evidence of inflammation[2].

Blood tests are not helpful in the diagnosis of CNS vasculitis. Cerebrospinal fluid (CSF) analysis is usually abnonnal in patients with PACNS, showing pleocytosis and elevated protein levels.The most important diagnostic aid remains the MRI and MRA. Brain biopsy is considered gold standard for confirming diagnosis and starting prolonged immunosuppressive therapy in this condition.[3] We did not do brain biopsy in this case because his parent did not give consent for it.

In PACNS, lesions on MRI are hyperintense on T2-weighted images and FLAIR sequences, and isointense or hypointense in T1-weighted images. Acute infarcts are represented by restricted diffusion on diffusion-weighted images (DWI).[4] MRA findings include vascular beading and absence or cut off of one or more vessels seen in single or multiple vascular beds.

Induction immunosuppression occurs over a 6-month period and includes 7 pulses of intravenous cyclophosphamide administered every 4 weeks, as well as high-dose corticosteroids.[5] Following the completion of induction with cyclophosphamide, maintenance therapy that consists of oral azathioprine or mycophenolate mofetil continues for 18 months, with weaning doses of oral corticosteroids. Methotrexate,

10-25 mg weekly, might be used in case of cyclophosphamide or azathioprine toxicity.

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