A CASE OF PRIMARY ANGIIITIS OF THE CENTRAL NERVOUS SYSTEM (PACNS)

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Abstract:
PACNS, a rare vasculitis limited to the brain, spinal cord and overlying leptomeninges. Atypical PACNS is the most common subset of PACNS. We report a 16 year old male who suffered young stroke (Right ganglio- capsular infarct) with headache. CT angiography showed occlusion of Rt.MCA segments. Takayasu arteritis, Fibromuscular dyplasia and PACNS were considered as differential diagnosis. Patient was diagnosed as Isolated CNS angiitis after excluding hypercoagulable state, infection and embolic etiologies. Patient was treated with corticosteroids for 5 months and repeat CT angiography showed persistence of the pre-existing lesion with no further progression. Hence, the case was diagnosed as an atypical subset of PACNS. This constitutes the group of CNS vasculitis which could not be entitled as granulomatous PACNS or benign angiopathy of CNS.

Keyword: Atypical angiitis, Primary CNS angiitis, young stroke. 16 years old male adolescent was admitted with the complaints of sudden onset of weakness in the left upper and lower limbs along with deviation of angle of mouth to right side while working out in gym. The weakness was associated with headache. No significant information regarding past medical, personal, family history was recorded. On examination, patient was conscious, oriented and afebrile. His blood pressure at the time of admission was recorded as 130/80 mm of Hg and the pulse rate was 89/min. His pulse was regular and felt in all peripheral vessels. Central nervous system examination revealed left sided hemiplegia with left upper motor neuron facial nerve palsy. No other focal neurological deficit was found. No abnormal findings were noted in other systems. The subsequent day plain CT scan of brain showed acute ischemic infarct in right ganglio capsular region and corona radiata. Chest X-ray and echocardiography were normal. Routine blood investigations which included ESR and CRP were normal. HIV antibody, Hbs antigen and anti-HCV were negative. Serum protein C and S, anti thrombin III, homocysteine levels were within normal limits. ANA, anti-cardiolipin antibody,
Lupus anticoagulant tests were negative. Fundus examination was normal. CSF analysis revealed no abnormalities. Hence, hypercoagulable state, embolic and infectious etiologies were ruled out. Since vasculitis was suspected, CT angiography of above arch and below arch vessels was performed. It showed complete occlusion of M1 and M2 segments of middle cerebral artery with distal reformation at M3 segment on the right side (Fig 1). All other arteries were normal. Renal artery doppler was normal.

1. CT cerebral angiography
Vasculitis like Takayasu arteritis, fibromuscular dysplasia and PACNS topped the list of differential diagnosis. The male gender, absence of string-of-beads appearance in middle cerebral artery (commonest finding in fibromuscular dysplasia) and absence of renal artery involvement were against the diagnosis of fibromuscular dysplasia. Male gender, absence of constitutional symptoms, inflammatory markers and other large and medium vessel involvement were against the diagnosis of Takayasu arteritis. Young age, absence of inflammatory markers and classical angiographic findings (cut off of M1 and M2 segments of middle cerebral artery) without other evidence of systemic vasculitis favours the diagnosis of PACNS. Patient was put on glucocorticoids (oral prednisolone 1mg/kg/day), then tapered gradually and maintained with low dose (oral prednisolone 10mg/day). Patient recovered well, but mild residual upper limb weakness persisted. Repeat cerebral angiography was performed after five months (fig.2). It showed persistence of the old lesion without any evidence of new lesions. Considering the acute onset of the symptoms, without any resolution of occluded artery segments in the subsequent angiography favoured the diagnosis of atypical PACNS. Atypical PACNS is the most common subset of PACNS.

DISCUSSION:
PACNS is a vasculitis limited to the brain, spinal cord and the overlying leptomeninges. There are several recognised clinical subsets. The modern era of PACNS began in the late 1950s. In 1988, Calabrese and Mallek proposed three working criteria for the diagnosis of PACNS1. 1.H/O unexplained neurological deficit that remains after a vigorous diagnostic workup including lumbar puncture and neurological imaging. 2.Either classic angiographic evidence (high probability) of vasculitis or histopathologic demonstration of vasculitis within the CNS.

Fig 2. CT cerebral angiography after 5 months
3. No evidence of systemic vasculitis or any other condition to which the angiographic or pathologic evidence could be attributed. The clinical subsets are granulomatous angiitis of CNS (GACNS), atypical PACNS, variants of PACNS which includes reversible vasoconstrictive disease states (RVDS), PACNS associated Varicella-Zoster infection and CNS sarcoid vasculitis. RVDS is otherwise called as benign angiopathy of CNS (BACNS).

A majority of patients (80%) with either angiographically or histopathologically documented PACNS do not fall neatly within the diagnostic categories of GACNS or BACNS. They are entitled as atypical PACNS. Some patients present with clinical features suggestive of BACNS, but their mode of presentation (subacute) along with abnormal spinal fluid (elevated WBC, protein) votes against BACNS. Some patients may present as RVDS like state but their repeat angiography fail to show dynamic improvement. Some patients may develop unusual anatomic presentations like spinal cord involvement. On neuroimaging enhancement within the cord like infiltrating appearance have been documented. A very few patients may present with mass lesion in the CNS. Differential diagnosis of PACNS is extensive and complex. Blood tests do not provide any valuable information to arrive at a diagnosis. For subacute presentation, the differential diagnosis of infections, neoplasms, inflammatory diseases require extensive laboratory testing, sampling of CSF and ultimately a biopsy of CNS. For acute presentation, an exhaustive search for emboli and hypercoagulability is essential. If none is found, cerebral angiography is recommended. Neuroimaging studies such as CT and MRI may show unilateral, multiple infarcts in the cortex, deep white matter or leptomeninges. The most important admonition in the diagnosis of CNS vasculitis is the fact that most patients will have a disease other than arthritis. So, the thoughtful differential diagnosis with the appropriate use of cerebral angiography and proper exclusionary diagnosis is mandatory to clinch the diagnosis. Steroids are the main modality of treatment.

REFERENCES:


