Relapsing myelopathy as the initial manifestation of primary central nervous system angiitis

Sir,

Primary angiitis of the central nervous system (PACNS) is a rare form of vasculitis exclusively affecting the CNS, without association with systemic processes. Symptomatic spinal cord involvement is infrequent and relapsing myelopathy as a presenting manifestation is extremely rare. We report a biopsy-proven case of PACNS, presenting with relapsing myelopathy as a sole manifestation for nearly 15 years that closely simulated multiple sclerosis.

A 37-year-old man was brought to our Emergency Department on 24th of June 2004 with acute onset of right hemiplegia and aphasia. He also had right focal motor seizures. His vital signs, including blood pressure was normal. He was drowsy and had global aphasia with dense right hemiplegia. Examination of other systems was unremarkable.

He gave a history of recurrent episodes of paraplegia starting from the age of 15 years. In September 1981, he was hospitalized for acute paraplegia with bladder retention and was diagnosed to have transverse myelitis at D12 spinal level. Cerebrospinal fluid (CSF) examination then was reported normal except for marginally raised protein of 55mg%. He recovered completely in three months with a short course of steroids and physiotherapy. In 1989 he had a similar episode of paraplegia and was treated at the same center.
with steroids. This time he had partial recovery, with residual bladder symptoms.

In November 1997, he had another episode of paraplegia. During this period he also had three episodes of seizures- two generalized and one left focal motor type of seizures. He was investigated with an MRI spine that showed intrinsic cord signal changes at D8-10 level, hyperintense on T1W and T2W images, suggestive of gliosis/infarct [Figure 1A]. T2W sequences of the MRI brain revealed multiple small hyperintense lesions in the subcortical white matter of the right frontal lobe and corpus callosum, reported as probable demyelination [Figure 1B]. A spinal angiogram to rule out a spinal AVM was normal. Workup for systemic vasculitis was negative. Based on a clinical suspicion of multiple sclerosis he was treated with intravenous methylprednisolone, 1gm/day for three days. Minimal lower limb weakness persisted with significant bladder symptoms.

During the present admission (June 2004), an emergency CT scan of brain revealed massive intracerebral hemorrhage involving the left parietal and frontal region with mass effect and intraventricular extension [Figure 1C]. Routine blood investigations including complete blood count, erythrocyte sedimentation rate, liver function tests, renal function tests were normal. Hemostatic parameters like bleeding time, clotting time, prothrombin time and activated partial thromboplastin time were normal. A cerebral angiogram showed luminal narrowing with alternating ectasia involving the cortical branches of the middle cerebral artery (MCA) bilaterally, suggestive of vasculitis [Figure 1D].

Patient deteriorated 24h after admission and developed minimal pupillary asymmetry. He underwent emergency craniotomy and evacuation of hematoma. A left frontal cortex biopsy was taken in the same sitting. The biopsy included the cortex with overlying meninges which showed inflammation in the subarachnoid space. Many of the leptomeningeal veins and venules coursing through the subarachnoid space were cuffed by tight clusters of well-formed epitheloid granulomas admixed with lymphocytes and small multinucleate giant cells [Figure 2A]. Extension of inflammatory process along the Virchow Robin space to surround parenchymal vessels produced perivascular non-necrotizing granulomas around parenchymal vessels [Figure 2B]. The adjacent cortex and subcortical white matter had small ischemic zones with reactive astrocysitis and gliosis in the subpial zone. These histological features were characteristic of granulomatous angiitis.

A detailed workup for secondary causes of granulomatous angiitis including RA factor, antinuclear antibody (ANA), cANCA, pANCA, HBsAg and anti-phospholipid antibodies, serum ACE level and CT scan of thorax for systemic vasculitis, sarcoidosis and chronic meningitis were all negative. The CSF VDRL, STPHA and HIV were all negative. A CSF analysis carried out eight weeks later showed no cells, with normal sugar and protein levels. He was started on steroids and cyclophosphamide pulse therapy. His recovery was partial with aphasia and residual right hemiparesis after six months of therapy.

Symptomatic involvement of the spinal cord has been documented much less frequently in PACNS than intracranial involvement. Most previous reports of isolated CNS vasculitis reported subacute progressive myelopathy spanning over months as a manifestation.\[5,6\] Acute transverse myelitis and relapsing myelopathy are rare manifestations of PACNS.\[4\] Rawlinson and Braun described a 19-year-old patient with relapsing myelopathy secondary to PACNS and reviewed four other patients with similar presentations.\[4\] But in all these patients the duration of symptoms was short.

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**Figure 1:** A. MRI T2W image of spinal cord done during last episode of myelopathy showing intramedullary hyperintense signal at D10-D11 level suggestive of cord infarct. B. MRI T2W image of brain showing subtle hyperintense signals in corpus callosum (arrows) and subcortical white matter. C. CT scan head plain at presentation showing intracerebral haemorrhage involving left parietal region with intraventricular extension and mass effect. D. Cerebral angiogram showing luminal narrowing with alternating ectasia involving cortical branches of both MCA.

**Figure 2:** A. Non-necrotizing epitheloid cell granulomas with giant cells (arrow) adjoining a leptomeningeal vessel (V). (H and E, x120). B. Thin walled vein (V) within the brain parenchyma shows epitheloid granulomas with giant cells destroying the wall. (H and E, x120)
Our patient had relapsing myelopathy as the sole manifestation for almost 15 years before he developed seizures and the final diagnosis was established only when he developed intracerebral hemorrhage 24 years after the onset of symptoms. This raises questions about the natural history of the disease and further widens the clinical manifestations associated with this disorder.

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References


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Letters to Editor

Spinal cord involvement and ganglionitis in leprosy

Sir,

We report an unusual case of leprosy to record the MRI changes of leprous ganglionitis and to newly document spinal cord involvement in leprosy,

A 20-year-old male presented with progressive loss of sensations and weakness of left hand of eight months duration. Examination showed a hypo-aesthetic patch on the dorsal aspect of the left elbow and left ulnar neuropathy involving sensory and motor functions. Left ulnar nerve was thickened at the elbow. A diagnosis of leprosy was made on clinical basis and he was started on Clofazimine, Rifampicin and Dapsone. He took treatment for three months and defaulted.

Two months after stopping therapy, he developed lightning pains affecting the medial aspect of the left arm and nape of neck. His clinical examination revealed involvement of the left ulnar and superficial branch of the left radial nerves, with loss of all sensory modalities including kinesthetic sensations. The electrophysiological findings are summarized in Table 1.

Biopsy of the left superficial radial cutaneous nerve demonstrated acid-fast lepra bacilli by Fite Faraco Stain [Figure 1]. The MRI of the cervical spine showed abnormality within the left half of cervical cord at C5-

Figure 1: Positive Fite Foracco stain for acid fast bacilli in nerve biopsy specimen

Figure 2: (A) T1W post contrast axial image of cervical spine at C5-6 showing evidence of ganglionitis and spinal cord changes and (B) T1W post contrast sagittal MRI of cervical spine showing intra medullary hyperintensity with cord swelling

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