SLE (systemic lupus erythematosus) is a multisystem autoimmune disorder of unknown aetiology which can present with myriad clinical presentations. The neurological manifestations of SLE consist of central nervous system (CNS) and peripheral nervous system manifestations (PNS). The CNS manifestations are aseptic meningitis, cerebrovascular accidents (stroke), demyelinating disorders, headache, involuntary movements like chorea, myelopathy, acute confusional states, cognitive dysfunction, mood disorder, seizures, psychosis and cranial nerve palsies.1 The PNS manifestations are Guillain Barre syndrome (GBS), autonomic disorder, mononeuropathy, polyneuropathy and plexopathy.1 Neuropathy in SLE can be clinically classified as mononeuritis multiplex and symmetrical and asymmetrical polyneuropathy. Symmetrical polyneuropathy being the most commonly seen clinical entity amongst the neuropathies in SLE. The neuropathy can be slowly progressive or acute in onset. Electrophysiologically, neuropathy is classified as axonal neuropathy, small fibre neuropathy, demyelinating neuropathy, mixed axonal-demyelinating sensorimotor polyneuropathy and plexopathy. Axonal neuropathy is further divided into sensory, sensorimotor and mononeuritis multiplex. Demyelinating neuropathy can be of two types: acute inflammatory demyelinating polyneuropathy (AIDP) and sensory demyelinating polyneuropathy. Anecdotal case reports also suggest that CIDP can occur as part of SLE neuropathy.2

The pathogenesis of lupus neuropathy is likely to be due to multifocal processes such as vasculitis causing destructive changes in vasa nervorum, demyelination, immune complex deposition and antibody-mediated damage.3

The clinical presentation of SLE neuropathy is like any other cause of neuropathy. The sensory symptoms consist of tingling or numbness and pain; while motor symptoms consist of weakness of the muscles supplied by the particular nerve e.g., wrist drop or foot drop. On clinical examination, there are decreased sensations in the distribution of the affected nerve. Bilateral ankle jerks are absent in symmetrical polyneuropathy. However, contrary to this traditional teaching, it is worth mentioning here that asymptomatic or subclinical neuropathy exists which is picked up only on electrophysiological studies.4 As mentioned earlier, the clinical presentation of lupus is so variable that neuropathy can occur in isolation or it may occur as a part of multisystem involvement with nephritis, anaemia or thrombocytopenia. Neuropathy may be the initial manifestation of active lupus or it may occur anytime during the course of the disease.

Once the diagnosis of SLE neuropathy is clinically suspected, it has to be confirmed with nerve conduction studies. The role of nerve conduction study is very important in classifying the type of neuropathy as discussed above. In a study reported by Oomtia et al,5 the prevalence of SLE neuropathy has been reported as 5.9 per cent. An interesting finding in this study was the presence of small fibre neuropathy which otherwise is not included in the 1999 ACR NPSLE case definitions.6 So the authors have used the EFNS and PNS guidelines and have done skin biopsy as part of their study to diagnose small fiber neuropathy.7 The technique of punch skin biopsy is a minimally invasive procedure to diagnose small fibre neuropathy.8 Small fibre neuropathy may be present despite normal electrophysiological studies i.e. when nerve conduction study is normal. In such patients punch skin biopsy can be very useful; particularly in patients who have small fibre neuropathy due to DRG neuronal loss. Since tissue diagnosis is an important tool in the era of evidence based medicine, the role of nerve biopsy still holds its place. The sural nerve is the most commonly biopsied nerve in establishing the diagnosis of neuropathy. So now, we have nerve biopsy and skin biopsy which can be used to arrive at a tissue diagnosis in case of SLE neuropathy.

Small fibre neuropathy can be of two clinicopathological entities affecting the dorsal root ganglions (DRGs) or the distal axons. The neuropathic pain can be length-dependent or non-length-dependent. Length-dependent neuropathic patients typically complain of symmetrical distribution of pain in distal...
lower limbs. But, the non-length dependent patients complain of pain which can affect the hands, face, torso, as well as proximal extremities simultaneously alongwith the distal lower limbs. In the study by Oomatia discussed above, it was also found that non-length-dependent small fibre neuropathy correlates with DRG neuronal loss, while non-length dependent small fibre neuropathy has distalmost axonal degeneration.

The prevalence of SLE neuropathy varies from 2-28 per cent. In a recent study reported by Xianbin W from China, the prevalence of SLE neuropathy is reported as1.5 per cent. IgG elevation has been attributed as a risk factor for developing peripheral neuropathy in these patients. A study from Malaya, reports the prevalence as 15.3 per cent. Florica et al have reported prevalence as 14 per cent. There are individual case reports which have reported lower motor neuron facial palsy. A case control study by Gaber et al have demonstrated diminished visual evoked potentials (VEP) and diminished blink reflexes (BR) to suggest cranial neuropathies also exist in SLE patients. In this study, they found that the abnormalities in VEP and BR were associated with antiribosomal P antibodies, hence they can be useful for detecting asymptomatic cranial neuropathy. Isolated case report of successful treatment of CIDP in SLE with steroids and cyclophosphamide is also found in published literature.

SLE neuropathy is a distinct entity which can respond to therapy as compared to the other aetiologies of peripheral neuropathy (where the patient may not improve very significantly). Immunosuppresants and corticosteroids are the mainstay of treatment. Pulse therapy with methylprednisolone and cyclophosphamide is very effective in the treatment of SLE neuropathy. Supportive care is given in the form of amitryptiline, nor-tryptiline, pregabalin, methylcobalamine, thiamine, pyridoxine and alpha-lipoic acids. The role of physiotherapy cannot be overemphasised. It is needless to say that demyelination and vasculitic neuropathy respond well as compared to axonal loss. Thus, once the diagnosis of SLE neuropathy is established, the further management is a teamwork which should consist of a rheumatologist, neurologist, dermatologist, physiotherapist, occupational therapist and primary care physician.

In the present article of JAPI, Saigal et al have reported SLE neuropathy to be 36% in their study population. The authors claim this to be higher than previous studies as they have done electrophysiological studies even in the asymptomatic patients. Vasculitic neuropathy is the commonest cause of neuropathy in this study. Apart from this, there is paucity of published literature from India about SLE neuropathy. I am sure readers of this issue will get stimulated after reading this article and we will have much more data from India in this field in subsequent years to come.

References