SYSTEMIC LUPUS ERYTHEMATOSIS HERALDING AS NEUROPATHY

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Abstract:
Systemic lupus erythematosus is associated with varied neurological manifestations. We are presenting SLE cases heralding with isolated peripheral nerve manifestations.

Methods:
A case series of 4 patients with rarer neuropathic features. One with AIDP, another with recurrent AIDP, another with severe axonal neuropathy, another with isolated small fibre neuropathy. All these patients on evaluation detected to have ANA dsDNA positivity and diagnosed to be SLE.

Discussion:
All the patients had only neuropathic manifestations. Neuropathic manifestations in SLE is seen in 4-27% of cases. Pathologically it can be vasculitic or due to autoimmune neuronal damage. The involvement of the peripheral nervous system in SLE is rare. It occurs in three forms, more common is distal symmetric sensory motor axonal poly neuropathy, which usually occurs late in the disease. Rare cases of a severe motor axonal neuropathy, mononeuritis multiplex and inflammatory demyelinating neuropathy are described in literature and can be the first symptom of SLE. Peripheral neuropathy appeared at the onset of the autoimmune disease in 50 of the patients. In the series presented all the four patients had neuropathy as heralding event. In cases of polyradiculoneuropathy without any possible preceding infectious cause or with recurrence, possibility of autoimmune disease should be ruled out. Conclusion: Peripheral nervous system involvement can be isolated and heralding manifestation of SLE manifesting in common or in rarer forms. Autoimmune disease evaluation for early detection should be part in patients with polyradiculoneuropathy not attributable to post infectious cause. These patients need early immunomodulation to prevent disease progression.

Keyword:
"SLE" "neuropathy" "small fibre" "demyelinating" "polyradiculopathy" Systemic lupus erythematosus heralding as neuropathy
Introduction:
Systemic lupus erythematosis is associated with varied neurological manifestations. In this, four cases of SLE are being presented, who had presented with rare isolated peripheral nerve involvement as the heralding event. Materials: This is a presentation of a case series of four patients with neuropathy features, on evaluation detected to have ANA/dsDNA positivity and diagnosed to have SLE.

Case 1: A 32 year old male presented with two episodes (5 years apart) of acute onset lower limb weakness ascended to flaccid quadriparesis in a week time. There was no wasting. CSF analysis showed high protein and normal other values. Nerve conduction studies showed demyelinating radiculoneuropathy in motor nerves in both episodes with normal sensory potentials. Patient had recovered completely with steroids in 2 weeks time in first episode and in one week time with plasma exchange in the second episode.

Case 2: A 28 year old female had acute areflexic flaccid quadriparesis with dysesthesias. CSF analysis showed proteins were elevated and normal other values. Nerve conduction studies showed demyelination pattern in motor and sensory nerves. Patient recovered completely in 8 weeks with plasma exchange and steroids.

Case 3: A 28 year old female presented with progressive ascending areflexic flaccid quadriparesis with wasting over 6 months. CSF analysis was normal. Nerve conduction studies showed axonopathic changes in motor nerves and normal SNAPs and a neurogenic pattern in Electromyogram. Biopsy showed multifocal axonopathy suggestive of vasculitis in the nerve and neurogenic atrophy in muscle. Patient had partial improvement with steroids, now being maintained on immunosuppressive medication.

Discussion:
The neuropathic manifestations in SLE is seen in 4-27%. Pathologically it can be vasculitic or autoimmune neuronal damage by anti neuronal antibody. Pathological data are supportive of a primary T-cell-mediated immune pathogenesis. The immediate cause of the vasculitic neuropathies is inflammation and occlusion of the vasa nervosum resulting in ischemia of the peripheral nerve. Immunoneuritis multiplex, the typical focal symptoms are thought to be caused by vasculitis related ischemic damage.

The involvement of the peripheral nervous system in SLE is rare. It occurs in three forms, more common is distal symmetric sensory motor axonal polyneuropathy, which usually occurs late in the disease. Rare cases of a severe motor axonal neuropathy, mononeuritis multiplex and inflammatory demyelinating neuropathy are described in literature and can be the first symptom of SLE. Peripheral neuropathy appeared at the onset of the autoimmune disease in about 50% of the patients. AIDP and Recurrent AIDP presentation is rare in SLE patients and had been described in previous case reports. The first two patients, one monophasic and another recurrent AIDP had similar Presentation Bodi et al., in their study on sural nerve biopsy sample taken from a patient of SLE neuropathy, found that endoneurial immune complex deposition also plays an important role in the demyelinating process and axonal damage seen in peripheral neuropathy.
A severe motor axonal neuropathy due to vasculitis has been described. The third patient had similar presentation with progressive motor weakness and wasting without sensory features. Isolated small fibre involvement may be manifestation of SLE. The fourth patient had similar presentation with normal nerve conduction studies and biopsy confirming vasculitic neuropathy with predominant small fibre loss.

Patients with SLE may have neuropathic symptoms despite normal nerve conduction studies and no clinical signs of peripheral neuropathy. In the literature, it has been mentioned that there is an abnormal reduction in intraepidermal small-diameter nerve fibre densities (IENF) in some patients despite normal function of their larger nerve fibers. This further supports the theory that a pure small-diameter nerve fibre neuropathy may occur in SLE. In patients with SLE, specific immunoglobulin deposits on neural surfaces or a low-grade inflammation of small blood vessels with an activated endothelium (vasculopathy) may, alone or in combination with other factors, render the small-diameter nerve fibres more vulnerable than larger fibres, or result in apoptotic signals that may be deleterious to small-diameter nerve fibres. The measurement of intraepidermal nerve fibre densities in skin biopsy specimens is considered an objective and reproducible method for evaluation of small diameter nerve fibres. Positive neuropathic symptoms are present only when an active pathogenic process is taking place in the nerve fibres, and that negative neuropathic symptoms will be the main findings when the IENFs are severely affected or destroyed. All the patients had only neuropathic manifestations and all the four had neuropathy as heralding event of the disease. In cases of polyradiculoneuropathy without any possible preceding infectious cause or with recurrence, possibility of autoimmune disease should be ruled out. In patients with a GBS, not responding to intravenous gammaglobulins, lupus should be excluded. In the case series, 3 of the 4 patients presented with predominant motor symptoms. Their recovery despite of their ages and other described factors were reasonable, the first two was complete as there was only demyelinating with no axonal involvement electrophysiologically and the third one had partial improvement as there was severe axonal involvement. The fourth patient who had presented predominantly with small fibre sensory symptoms did not show improvement with scheduled immunomodulatory therapy suggesting that the extent of axonal involvement determines the recovery. In contrast to vasculitic neuropathy associated with systemic vasculitis, the prognosis was good with isolated vasculitic neuropathy. Prednisone and other immunosuppressive like cyclophosphamide should be considered in patients with GBS as a feature of lupus. Relapse occurred almost exclusively in patients treated with prednisolone alone. Aggressive early treatment with immunosuppressives like cyclophosphamide may prevent relapse.

Conclusion: Peripheral nervous system involvement can be the isolated and heralding manifestation of SLE manifesting in common or in rarer forms. Autoimmune disease evaluation for early detection should be a part in patients with polyradiculoneuropathy not attributable to post infectious cause. These patients need early immunomodulation to modify disease progression.
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