Commentary

Neuropsychiatric manifestation of systemic lupus erythematosus (SLE) is one of the chief and most detrimental presentations. It involves a wide range of neurological manifestations affecting the central, peripheral and autonomic nervous systems, as well as psychiatric syndromes. The clinical features are characterized by some researchers as either diffuse (e.g. encephalopathy, coma, depression, and psychosis) or complex (e.g. encephalopathy with stroke or seizure and psychiatric presentation with stroke or seizures). Neuropsychiatric manifestations are present in ≈ 50-70% of patients with SLE. However, cerebellar ataxia is rarely reported (<2%).[1-3] The cause of the cerebellar ataxia in SLE case report in the current issue {Ghosh K, Chatterjee A, Ghosh S, Chakraborty S. Cerebellar ataxia in a young patient: A rare path to lupus. Journal of Neurosciences in Rural Practice}[4] was cerebellar atrophy, other reported causes, including cerebral infarction, antineuronal antibody and vasculopathy.[1,3]

The diverse nature of CNS syndromes in SLE proposes that different disease mechanisms may be involved. An association between neuropsychiatric symptoms and specific antibodies has been demonstrated in a few studies.[5-7] Deposition of immune complexes in the choroid plexus, the effects of antibodies; anticardiolipin and anti-neuronal antibodies cause neural cell dysfunction and immune-mediated vascular disease.[5-7] Lupus anticoagulant and anticardiolipin antibody have been found to be associated with various neurological manifestations in SLE patients. In addition to thrombotic events, antiphospholipid antibodies have been shown to cross-react with epitopes on CNS phospholipids like sphingomyelin, causing neuronal dysfunction.[5,7,8] Embolic disease secondary to Libman-Sachs endocarditis and large-vessel vasculitis have also been described.[6]

Although it would be necessary to have diagnostic tests to establish the diagnosis of neuropsychiatric lupus, such tests do not exist. Thus, the approach to lupus patients with neuropsychiatric manifestations, like the lupus case report with cerebellar ataxia[4] consists of confirmation of the diagnosis of SLE. Serologic tests are used to establish the diagnosis of SLE. The antinuclear antibody titer is positive in almost all patients. Serum levels of anti-DNA and anti-Sm antibodies and low levels of complement are useful in the diagnosis of SLE.[1,2] Other serologic tests include estimation of anticardiolipin, anti-neuronal and antiphospholipid antibodies. The next step is to distinguish between organic and functional etiologies of ataxia and exclude other disorders involving the cerebellum e.g. acute disseminated encephalomyelitis, diffusely infiltrating glioma or lymphoma and drug-related inflammatory processes.[5,9] MRI is the most useful neuroimaging study in patients with SLE, particularly in those with focal neurologic deficits. It is the modality of choice to demonstrate cerebellar pathology, which may be undetected on CT brain. MRI is highly useful in the diagnosis of acute cerebellar ataxia.[9]

The management SLE patients with CNS involvement depends on immunosuppressive therapies with azathioprine, cyclophosphamide, mycophenoate mofetil and rituximab in association with corticosteroids (oral or intravenous pulse therapy).[5]

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