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Neurosurgical Approaches in Demyelinating Disorders; Where are We Now?

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Demyelinating diseases of the central nervous system (CNS) are a group of autoimmune disorders affecting the myelin sheets of CNS neurons resulting in different neurological deficits and disability. Multiple sclerosis (MS), Devic's disease, progressive multifocal leukoencephalopathy, acute disseminated encephalomyelitis (ADEM), and neuromyelitis optica (NMO) are among the most common types of demyelinating disorders [1]. Currently MS is the leading cause of neurological disability in young population after trauma [2]. The incidence and prevalence of MS is increasing worldwide primarily due to increase the incidence in female population [3]. Globally, the median estimated incidence of MS is 5.2 (range: 0.5-20.6) per 100,000 p-yrs, the median estimated prevalence of MS is 112.0 (with a range of 5.2-335) per 100,000 p-yrs, and the average disease duration is 20.2 years (range: 7.6-36.2) [4]. In Iran, the prevalence and incidence of MS is estimated to be 54.51 and 5.87 per 100,000 [5, 6]. Diagnosis of demyelinating disorders is a controversial issue and several criteria has been introduced for the aforementioned subject. Diagnostic criteria for clinically definite MS require documentation of two or more episodes of symptoms and two or more signs that reflect pathology in anatomically noncontiguous white matter tracts of the CNS. The second may be documented by abnormal paraclinical tests such as MRI or evoked potentials (EPs) [7].

The standard treatment for patients with demyelinating disorders and especially MS is the medical management. Recent large placebo-controlled trials in relapsing-remitting multiple sclerosis have shown efficacy of new oral dis-

ease-modifying drugs, teriflunomide and dimethyl fumarate, with similar or better efficacy than the injectable disease-modifying drugs, IFN- β and glatiramer acetate. In addition, the new oral drugs seem to have a favorable safety profile. Further, the monoclonal antibody alemtuzumab, which in clinical trials has shown superiority to subcutaneous IFN- β 1a, has been approved in Europe [8]. In acute exacerbation and flare up of the disease, methylprednisolone pulse is the only approved treatment [9].

Although the standard of treatment of MS and other demyelinating disorders is medical, but neurosurgical procedures especially the functional neurosurgical interventions has found their way in management of patients with demyelinating disorders. These interventions are used to treat the complications of MS and demyelinating disorders such as tremor, trigeminal neuralgia, movement disorders and neuropathic pains. Most of these interventions are stereotactic in order to obtain precise targeting of a special deep brain nucleus [10]. The first application of deep brain stimulation (DBS) in patients with MS was for treatment of tremor. Recently it has been demonstrated that DBS of ventral intermediate (VIM) nucleus of thalamus results in alleviation of severe, disabling tremor in patients with MS [11]. Other experiments have also demonstrated decreased tremor and improved quality of life in MS patients responsive to DBS [10, 12].

Neuropathic pain and trigeminal neuralgia are among the other complications of demyelinating disorders which are associated with disability and decreased quality of life [2]. Several investigations have shown that DBS of thalamic

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nuclei would be effective in treatment of neuropathic pain in patients with MS, although the results are controversial and need further investigations [13]. The trigeminal neuralgia is often treated by microsurgical decompression of the trigeminal nerve. However this approach is not effective in patients with demyelinating disorders as they have different pathology. Case series have shown that Gamma Knife surgery (GKS) is an effective and safe treatment for trigeminal neuralgia in patients with MS [14]. No clinical trial is available for comparing the results and outcome between GKS and microsurgical approach. However results of case series are favorable.

There are still other field that could be approached surgically in patients with demye-

lating disorders. Recent bodies of evidence have demonstrated a link between cervical cord plaques and discopathy in those with MS [15] which needs further investigation. Tumefactive demyelinating lesions (TDL) are another issues observed in patients with MS which have unknown course and pathology [16]. The link between demyelinating disorders and brain tumors is also another issues which deserves further investigation [17]. To take a long story short, although the role of functional neurosurgical procedures in management of patients with demyelinating disorders especially MS is still limited, but there is optimistic horizons for growing role of neurosurgical procedures in management of different complications of demyelinating disorders refractory to medical therapy.

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