International Journal of Neurology Research
Online ISSN: 2664-9098, Print ISSN: 2664-908X
Received: 02-11-2018; Accepted: 03-12-2018; Published: 02-01-2019
www.neurologyjournals.com
Volume 1; Issue 1; 2019; Page No. 03-06

Diseases of the nervous system: A basic review

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Abstract
Nervous System (basic review) are to understand very easily for graduate, post graduate and post doctoral ayush, dental, medical etc., students. I am explaining main and important diseases in nervous system in day to day practical life for medical students and professionals. Diseases are Alzheimer's disease, Parkinson's disease, Gillian Barr's syndrome and myasthenia gravis.

Keywords: nervous system, causes, clinical features, investigation, management.

1. Introduction
We have lot of diseases to explain in nervous system. But only main/few diseases are reviewing for under graduate, post graduate and post-doctoral ayush, dental, medical, nursing etc., entrance and main examination purpose.

Alzheimer's disease
Alzheimer's disease exists along a spectrum, from early memory changes to functional dependence and death. Using a case illustration, we review the evaluation and diagnosis of mild cognitive impairment and the diagnosis and management of Alzheimer's disease at each stage, including the management of both cognitive and behavioral/psychiatric aspects of the disease and end-stage and end-of-life care. Dementia is a clinical syndrome characterized by progressive decline in two or more cognitive domains, including memory, language, executive and visuospatial function, personality, and behavior, which causes loss of abilities to perform instrumental and/or basic activities of daily living. Alzheimer's disease (AD) is by far the most common cause of dementia and accounts for up to 80% of all dementia diagnoses. Although the overall death rate in the United States from stroke and cardiovascular disease is decreasing, the proportion of deaths related to AD is going up, increasing by 89% between 2000 and 2014. Current treatments available include cholinesterase inhibitors for patients with any stage of AD dementia and memantine for people with moderate-to-severe AD dementia. These medications have been shown to enhance the quality of life for both patient and caregiver when prescribed at the appropriate time during the course of illness; however, they do not change the course of illness or the rate of decline [1].

Clinical features
Both short term and long term memory are affected, apraxia, visuo spatial impairment, anosognosia, depression, cannot identify person and the clinical features are made acutely worse by coexistent intercurrent illness [2].

Management
There is no specific treatment for this condition.

Parkinson's disease
Parkinson's disease (PD) was first described by Dr. James Parkinson in 1817 as a "shaking palsy." It is a chronic, progressive neurodegenerative disease characterized by both motor and nonmotor features. The disease has a significant clinical impact on patients, families, and caregivers through its progressive degenerative effects on mobility and muscle control. The motor symptoms of Parkinson's disease are attributed to the loss of striatal dopaminergic neurons, although the presence of nonmotor symptoms supports neuronal loss in nondopaminergic areas as well. The term Parkinsonism is a symptom complex used to describe the motor features of Parkinson's disease, which include resting tremor, bradykinesia, and muscular rigidity. Parkinson's disease is the most common cause of Parkinsonism, although a number of secondary causes also exist, including diseases that mimic Parkinson's disease and drug-induced causes. Parkinson's disease is one of the most common neurodegenerative disorders. The Parkinson's disease Foundation reports that approximately 1 million Americans currently have the disease. Although it is primarily a disease of the elderly, individuals have developed Parkinson's disease in their 30s and 40s. Gender differences pertaining to the incidence of Parkinson's disease are reflected in a 3:2 ratio of males to females, with a delayed onset in females attributed to the neuroprotective effects of estrogen on the nigrostriatal dopaminergic system. Parkinson's disease variable but pronounced progression has a significant impact on patients, families, and society. Advanced and end-stage disease may lead to serious complications, including pneumonia, which are often associated with death [3].
Parkinson's disease is a disorder of the extrapyramidal system, which includes motor structures of the basal ganglia, and is characterized by the loss of dopaminergic function and consequent diminished motor function, leading to clinical features of the disease. Progressive degeneration

of dopaminergic neurons in the substantia nigra pars compacta (SNpc), which project to the striatum (the nigrostriatal pathway), results in the loss of dopaminergic function in individuals with PD. Typically, patients experience the motor features of Parkinson's disease only after 50% to 80% of dopaminergic neurons have been lost, suggesting the involvement of a compensatory mechanism in the early stages of the disease. Two types of dopamine receptors, D_1 (excitatory type) and D_2 (inhibitory type), influence motor activity in the extrapyramidal system. Components of this system include the basal ganglia, which involves the internal globus pallidal segment (GPi) of the ventral striatum, and the pars reticulata portion of the substantia nigra (SNpr). These components are part of larger circuits located in the thalamus and the cortex. The loss of dopamine in the striatum of Parkinson's disease patients results in increased activity in the GPi/SNpr circuits and subsequent gamma aminobutyric acid (GABA) dysfunction, leading to inhibition of the thalamus. The end result is the decreased ability of the thalamus to activate the frontal cortex, resulting in the decreased motor activity characteristic of Parkinson's disease. Accordingly, restoring dopamine activity in the striatum through D_2 and D_1 receptor activation with dopaminergic therapies mediates clinical improvement in the motor symptoms of Parkinson's disease [4]. In addition, dopaminergic loss results not only in reduced activation of the thalamus but also in increased cholinergic activity due to the loss of dopamine's normal inhibitory influence.

Clinical features

Clinical features of Parkinson's disease are tremor, rigidity, bradykinesia, may be absent initially, when non-specific symptoms of tiredness, aching limbs, mental slowness, small handwriting may be noticed. Most patients have difficulty with rapid fine movements, tremor also affects the legs, mouth as well as tongue, slowness of gait difficulty with tasks such as fastening buttons, shaving or writing, postural righting reflexes are impaired early on in the disease, speech become softer, indistinct.

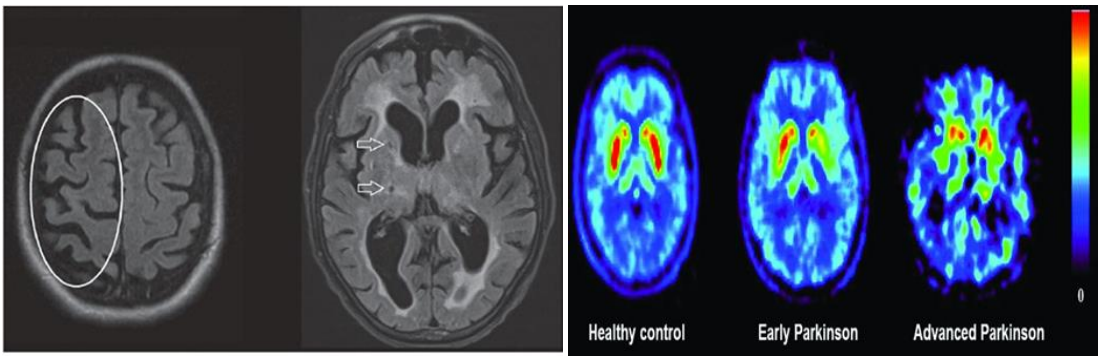

Fig 1: Parkinson's MRI brain & SPECT

Investigations

The differential diagnosis of PD should include a comprehensive history and physical examination. Difficult or questionable cases should be referred to a movement-disorder specialist for further evaluation. There are no definitive tests to confirm the diagnosis of PD; therefore, a clinical diagnosis requires the clinician to review the patient's history, to assess symptoms, and to rule out alternative diagnoses, such as multiple-system atrophy, DLB disease, and essential tremor. The cardinal motor features of PD—described as the "classical triad"—include a 4-Hz to 6-Hz resting tremor, "cogwheel" rigidity, and bradykinesia [5].

Management

The primary goal in the management of PD is to treat the symptomatic motor and nonmotor features of the disorder, with the objective of improving the patient's overall quality of life. Appropriate management requires an initial evaluation and diagnosis by a multidisciplinary team consisting of neurologists, primary care practitioners, nurses, physical therapists, social workers, and pharmacists. Effective management should include a combination of no-pharmacological and pharmacological strategies to maximize clinical outcomes. To date, therapies that slow the progression of PD or provide a neuroprotective effect have not been identified [6].

Guillain barre's syndrome [7-10].

It is acute inflammatory demyelinating polyneuropathy. Guillain Barré syndrome (GBS) was first described in 1916 (Guillain G, 1916) and is approaching its 100th anniversary. Our knowledge of the syndrome has hugely expanded since that time. Once originally considered to be only demyelinating in pathology us now recognizes both axonal and demyelinating subtypes. Numerous triggering or antecedent events including infections are recognized and GBS is considered an immunological response to these. GBS is now considered to be a clinical syndrome of an acute inflammatory neuropathy encompassing a number of subtypes with evidence of different immunological mechanisms. Some of these are clearly understood while others remain to be fully elucidated. Complement fixing antibodies against peripheral nerve gangliosides alone and in combination are increasingly recognised as an important mechanism of nerve damage. New antibodies against other nerve antigens such as neurofascin have been recently described. Research databases have been set up to look at factors associated with prognosis and the influence of intravenous immunoglobulin (IvIg) pharmacokinetics in therapy. Exciting new studies are in progress to examine a possible role for complement inhibition in the treatment of the syndrome.

Early studies reported oedema of the peripheral nerves with

Sparse inflammatory infiltrate. Classic studies by Asbury and colleagues emphasised the importance of perivascular lymphocytes which resembled the findings in the animal model experimental allergic neuritis.

Clinical features

Clinical features of Guillain barre's syndromes are muscle weakness, paraesthesial especially distal and limb pains often precede the weakness, facial or bulbar weakness, respiratory failure can develop within hours.

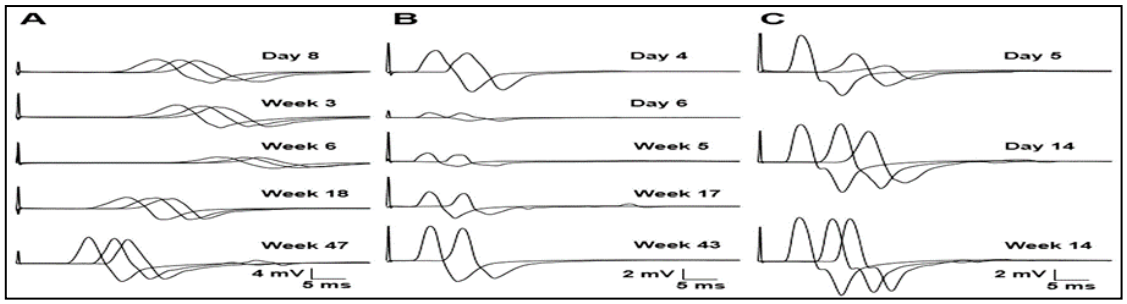

Fig 2: Guillain barre's syndrome electromyography

Diagnosis

A lumbar puncture should be done before treatment. A cerebrospinal fluid (CSF) white cell count of over 10/µl raises the possibility of leptomeningeal malignancy, HIV or an alternative infectious diagnosis (EG Lyme disease or poliomyelitis), but in clinical trials CSF cell counts up to 50/µl are permitted. IvIg can very occasionally cause aseptic meningitis. Typically, the CSF protein is raised after the first week, often to more than 1 g/l. Routine blood tests should include creatine kinase, biochemistry and Ig levels. These are done to exclude other causes of weakness and to reduce the risks of ivIg. In renal failure ivIg is relatively contraindicated and it is more likely to cause anaphylaxis in patients with IgA deficiency.

Management

Treatment should be started as soon as possible, but there is no evidence that starting it 12 hours earlier (eg overnight) makes any difference. First-line treatment is now usually ivIg because of its ease of administration. Adequate pain relief and a multidisciplinary approach to rehabilitation are essential, as is patient education during the slow but steady recovery, with improvements to be expected for up to two years.

Myasthenia gravis

It is characterized by progressive inability to sustain a maintained or repeated contraction of striated muscle (fatigability). Acquired myasthenia gravis is a relatively uncommon disorder, with prevalence rates that have increased to about 20 per 100,000 in the US population[11]. This autoimmune disease is characterized by muscle weakness that fluctuates, worsening with exertion, and improving with rest. In about two-thirds of the patients, the involvement of extrinsic ocular muscle presents as the initial symptom, usually progressing to involve other bulbar muscles and limb musculature, resulting in generalized myasthenia gravis. Although the cause of the disorder is unknown, the role of circulating antibodies directed against the nicotinic acetylcholine receptor in its pathogenesis is well established.

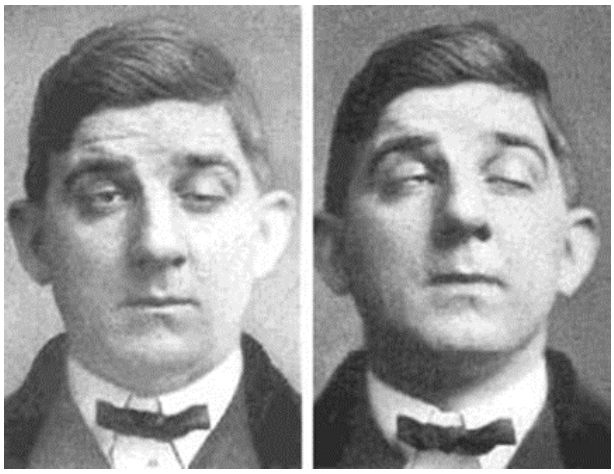

Fig 3: Myasthenia gravis

The nerve terminals innervating the neuromuscular junctions (NMJ) of skeletal muscles arise from the terminal arborization of α-motor neurons of the ventral horns of the spinal cord and brain stem. The NMJ itself consists of a synaptic cleft and a 20 nm thick space that contains acetylcholinesterase (ACHE) along with other supporting proteins/proteoglycans [12]. The NMJ postsynaptic membrane has deep folds with acetylcholine receptors (ACHR) tightly packed on the top of these folds. When the nerve action potential reaches the synaptic bouton, depolarization opens voltage gated Calcium channels on the presynaptic membrane, triggering release of ACh into the

synaptic cleft. The ACh diffuses into the synaptic cleft to reach postsynaptic membrane receptors where it triggers off the end-plate potential (EPP) and gets hydrolyzed by AChE within the synaptic cleft. MuSK (muscle specific tyrosine kinase), a postsynaptic transmembrane protein, forms part of the receptor for agrin, a protein present on synaptic basal lamina. Agrin/MuSK interaction triggers and maintains rapsyn-dependent clustering of AChR and other postsynaptic protein. Rapsyn, a peripheral membrane protein on the postsynaptic membrane, is necessary for the clustering of AChR. Mice lacking agrin or MuSK fail to form NMJs and die at birth due to profound muscle weakness [13, 14].

Clinical features

The disease usually presents between the age of 15 and 50 years with women affected more often than men. Symptoms are abnormal fatigable weakness of the muscles, movement is initially strong, it rapidly weakness, ptosis, diplopia, limb muscle may be affected, mostly commonly those of the shoulder girdle, respiratory muscles may be involved, aspiration may occur if the cough is ineffectual. Sudden weakness from a cholinergic or myasthenic crisis may require ventilator support [15].

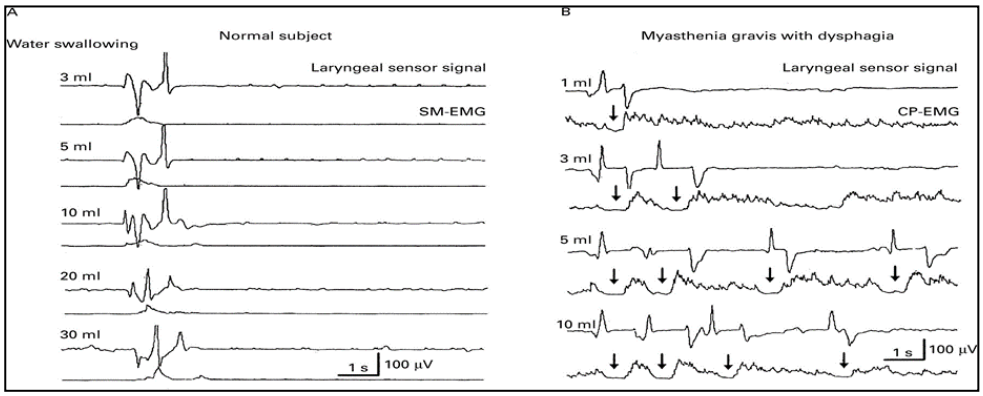

Fig 4: Myasthenia gravis electromyography

Diagnosis

For a patient with ptosis, a small cube of ice is placed over the eyelid for about 2 minutes. Improvement of the ptosis after this procedure suggests a disorder of neuromuscular transmission. all patients with MG should have a computed tomography (CT) scan of the chest done with contrast. Routine chest radiography may be done but should not be done in place of the CT scan of the chest[16].

Management

Thymectomy should be performed as soon as feasible in any patient with myasthenia not confined to extra ocular muscles, unless the disease has been established for more than 7 years. Plasma exchange are removing antibody from the blood may produce marked improvement.

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