Oral Medicine

Neuromuscular Disorders II

Lecture 14

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ALZHEIMER'S DISEASE (AD)

Dementia is defined as an acquired deterioration in cognitive abilities that impairs the successful performance of activities of daily living.

Memory is the most common cognitive ability lost with dementia; other mental faculties affected include problem-solving skills, judgment, visuospatial ability, and language.

The genetic basis of AD has been studied extensively, and specific genetic mutations have been implicated in both the familial and sporadic forms of the disease.

Familial AD is an autosomal dominant disorder with onset typically prior to age 65 year.

Clinical Manifestations

AD is a slowly progressive disorder represented by a continuum recognizes three stages of AD:
(1) Preclinical AD (2) Mild cognitive impairment due to AD (3) Dementia due to AD.

Preclinical AD occurs before changes in cognition, and everyday activities are observed and primarily used for research purposes. (During this phase, the condition is asymptomatic but detectable on a screening test).

Cognitive impairment (CI) due to AD is characterized by mild changes in memory and other cognitive abilities that are noticeable to patients and families but are not sufficient to interfere with day to-day activities.

Dementia due to AD is characterized by changes in two or more aspects of cognition and behaviour that interfere with the ability to function in everyday life.
The initial signs of AD involve retrograde amnesia from progressive declines in episodic memory. This may initially go unrecognized or be viewed; however, as the disease progresses, memory loss begins to affect performance of daily activities, including following instructions, driving, and normal decision making.

As AD progresses, the individual is often unable to work, gets confused and lost easily, and may require daily supervision, language impairment and loss of abstract reasoning skills.

Advanced AD is characterized by loss of cognitive abilities, agitation, delusions, and psychotic behavior.

Patients may develop muscle rigidity associated with gait disturbances and often wander aimlessly.

In end-stage AD, patients often become rigid, mute, incontinent, and bedridden. Help is needed for basic functions, such as eating and dressing, and patients may experience generalized seizure activity.

Death often results from malnutrition, heart disease, pulmonary emboli, or secondary infections.

Diagnosis

Diagnosis of preclinical AD primarily utilizes biomarker assessment, including markers of Aβ protein deposition in the brain, and markers of downstream neurodegeneration (elevated CSF tau protein) and brain atrophy on MRI.

Clinical diagnosis of AD is based on an individual's medical history together with the clinical and neurologic examination findings.

Criteria include a history of progressive deterioration in cognitive ability in the absence of other known neurologic or medical problems.

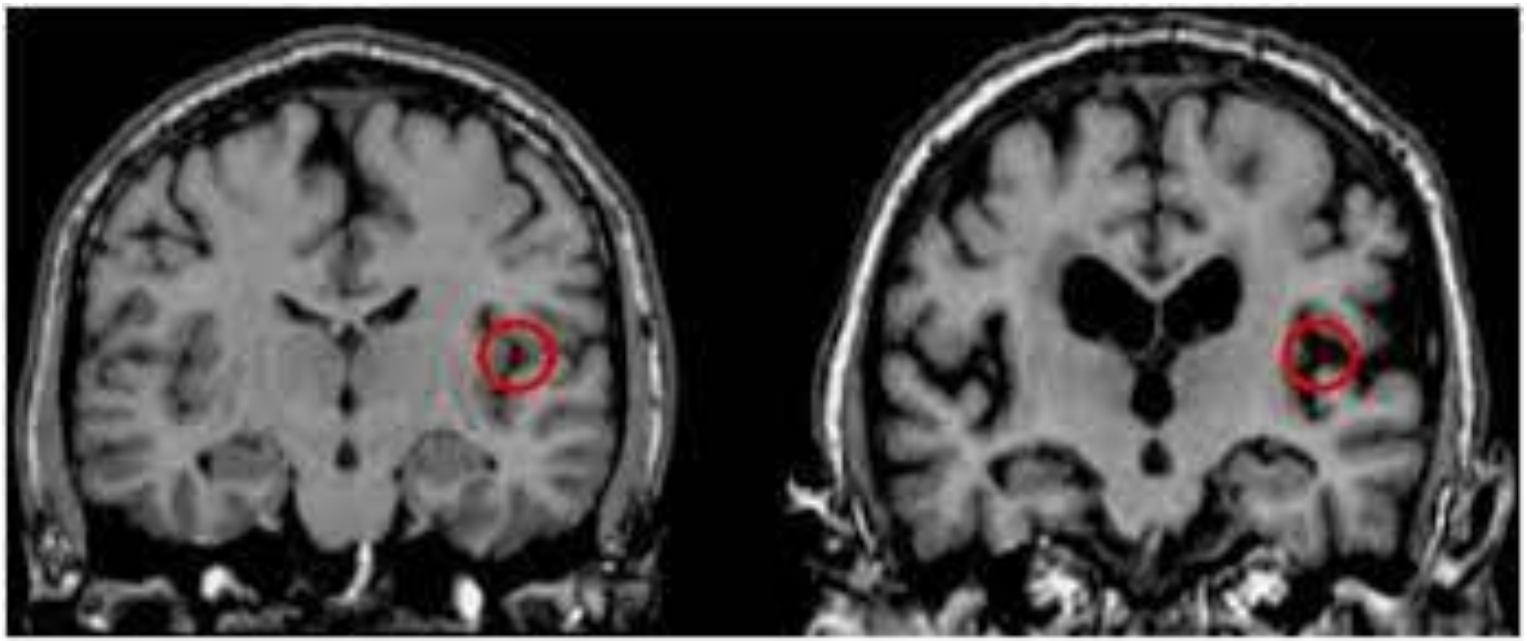
Possible AD refers to those who meet the criteria for dementia but have another illness that may contribute to the neurologic status, such as hypothyroidism or cerebrovascular disease, vitamin deficiency, depression, delirium, side effects of drugs and toxicity and excessive use of alcohol .

Diagnostic analysis of CSF may show a slight increase in tau protein and a lower concentration of Aβ peptide compared with healthy individuals or those with other dementias.

Electroencephalographic (EEG) studies typically demonstrate generalized slowing without focal features.

Neuroimaging is important in evaluating suspected AD to exclude alternative causes of dementia, such as cerebrovascular disease, subdural hematoma, or brain tumor.

MRI and CT typically reveal dilatation of the lateral ventricles and widening of the cortical sulci, particularly in the temporal regions.



Treatment

There is no cure for AD, and therapy is aimed at slowing the progression of the disease. **Cholinesterase inhibitors** are approved to treat mild to moderate cases of AD and are considered the standard of care.

Memantine, a noncompetitive *N*-methyl-d-aspartate receptor antagonist believed to protect neurons from glutamate-mediated excitotoxicity, is used for treatment of moderate to severe AD.

Studies have demonstrated greater cognitive and functional improvement when memantine is used in conjunction with cholinesterase inhibitors compared to monotherapy.

Antidepressants, such as selective serotonin reuptake inhibitors, are commonly used to treat depression, which is often seen in the mild to moderate stages of AD.

Antipsychotic agents are used for those patients who display aggressive behavior and psychosis, especially in the later stages of the disease.

Other agents that have been reported to be of clinical value in the treatment of AD include: antioxidants such as α-tocopherol (vitamin E) and cholesterol-lowering drugs.

Oral Health Considerations

Oral and dental health is a major issue in patients with AD because significant deterioration in oral health status is commonly observed with advancing disease.

Patients with AD appear to be at higher risk for developing coronal and root caries, periodontal infections, TMJ abnormalities, and orofacial pain compared to healthy subjects.

Patients with AD should be placed on an aggressive preventive dentistry program, including an oral examination, oral hygiene education, prosthesis adjustment and a three-month recall.

It is recommended to complete restoration of oral health-care function in the earliest stages of AD because the patient's ability to cooperate diminishes as cognitive function declines.

Time-consuming and complex dental treatment should be avoided in persons with severe AD.

Medications used to treat AD can cause a variety of orofacial reactions and potentially interact with drugs commonly used in dentistry.

Cholinesterase inhibitors may cause sialorrhea, whereas antidepressants and antipsychotics are often associated with xerostomia.

In addition, dysgeusia and stomatitis have been reported with use of antipsychotic agents.

Antimicrobials such as clarithromycin, erythromycin and ketoconazole may significantly impair the metabolism of galantamine resulting in central or peripheral cholinergic effects.

Anticholinesterases may increase the possibility of gastrointestinal irritation and bleeding when used concomitantly with NSAIDs.

Local anaesthetics with adrenergic vasoconstrictors should be used with caution in AD patients taking tricyclic antidepressants due to potential risk of cardiovascular effects such as hypertensive events or dysrhythmias.

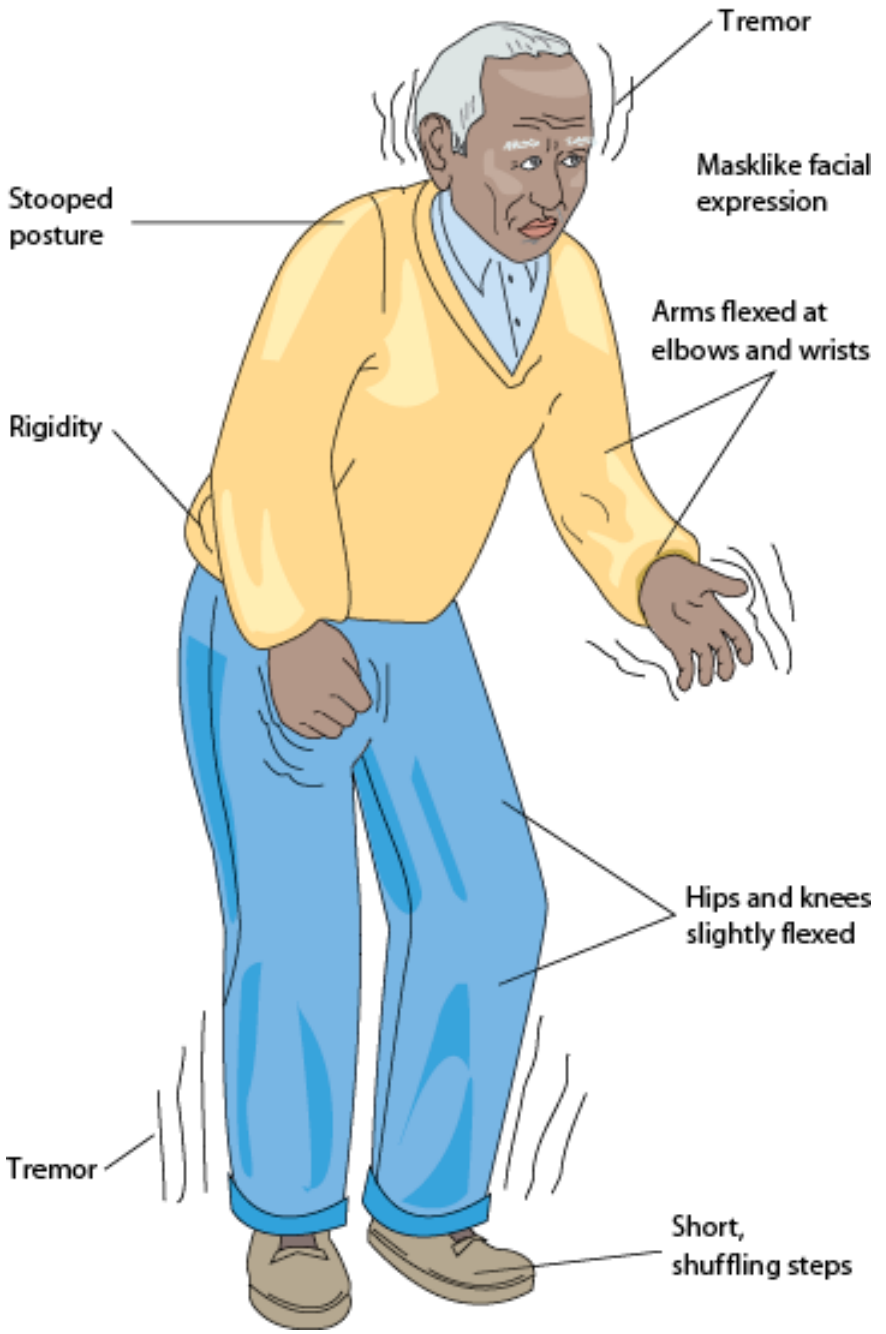
Parkinsonism

Is an idiopathic neurodegenerative disorder that mainly affects adult in middle or late life.

It is due to dopamine depletion in the brainstem lead to an imbalance of dopamine and acetylcholine (neurotransmitters that are normally present in the corpus striatum).

The cardinal features of parkinsonism include:

1- Rigidity
2- Tremors
3- Bradykinesia
4- Postural instability.



Many of the signs of Parkinson’s disease are found in the head and neck:

- The typical “mask like” facial appearance with infrequent blinking and lack of expression is caused by bradykinesia.
- The muscle rigidity also causes difficulty in swallowing, resulting in saliva drooling.
- Speech affected because of the lack of muscle control, and mandibular tremor results in masticatory difficulties, especially in those with removable dental appliances.
- Abnormalities in oral behavior such as purposeless chewing, grinding and sucking movements, are also well recognized in patients with Parkinson’s disease and make dental treatment especially difficult.

Treatment

Drug treatment is often not required early in the course of parkinsonism.

Levodopa, a dopamine precursor that can cross the blood-brain barrier, improves all the major features of parkinsonism.

Bell's palsy

It is recognized as a unilateral paresis of the facial nerve. It has been attributed to an inflammatory reaction involving the facial nerve. (compression of the facial nerve in it's canal)

A relationship has been demonstrated between Bell’s palsy and the isolation of herpes simplex virus 1 from nerve tissues.

- Bell’s palsy begins with slight pain around one ear, followed by an abrupt paralysis of the muscles on that side of the face.
- The eye on the affected side stays open, the corner of the mouth drops, and there is drooling.
- As a result of masseter weakness, food is retained in both the upper and lower buccal and labial folds.
- The facial expression changes remarkably, and the creases of forehead are flattened.
- Due to impaired blinking, corneal ulcerations from foreign bodies can occur.

Diagnosis

There's no specific test for Bell's palsy. Look at face and ask to move facial muscles by wrinkled forehead and lifting brow, closing eyes, showing teeth and blow out his cheeks.

Crease up the forehead

Keep eyes closed against resistance

Reveal the teeth

Puff out the cheeks

Other conditions (such as a stroke, Lyme disease and tumors) can also cause facial muscle weakness, mimicking Bell's palsy, may recommend other tests, including:

- **Electromyography (EMG):** This test can confirm the presence of nerve damage and determine its severity. An EMG measures the electrical activity of a muscle in response to stimulation and the nature and speed of the conduction of electrical impulses along a nerve.

- **Imaging scans:** MRI or CT may be needed on occasion to rule out other possible sources of pressure on the facial nerve, such as a tumor or skull fracture.

Treatment

Commonly used medications to treat Bell's palsy include:

Corticosteroids, such as prednisone is a powerful anti-inflammatory agent. That can reduce the swelling of the facial nerve, so it will fit more comfortably within the bony corridor that surrounds it.

Antiviral drugs: the role of antivirals remains unsettled. Antivirals alone have shown no benefit compared with placebo.

However, when antivirals added to steroids may show decrease the severity and length of paralysis.

Valacyclovir (Valtrex) is sometimes given in combination with prednisone in people with severe facial palsy.
It is also helpful to protect the eye with lubricating drops or ointment and a patch if eye closure is not possible.

Physical therapy:

Paralyzed muscles can shrink and shorten, causing permanent contractures.
A physical therapy by massage and exercise of facial muscles to help prevent this from occurring.

Surgery:-

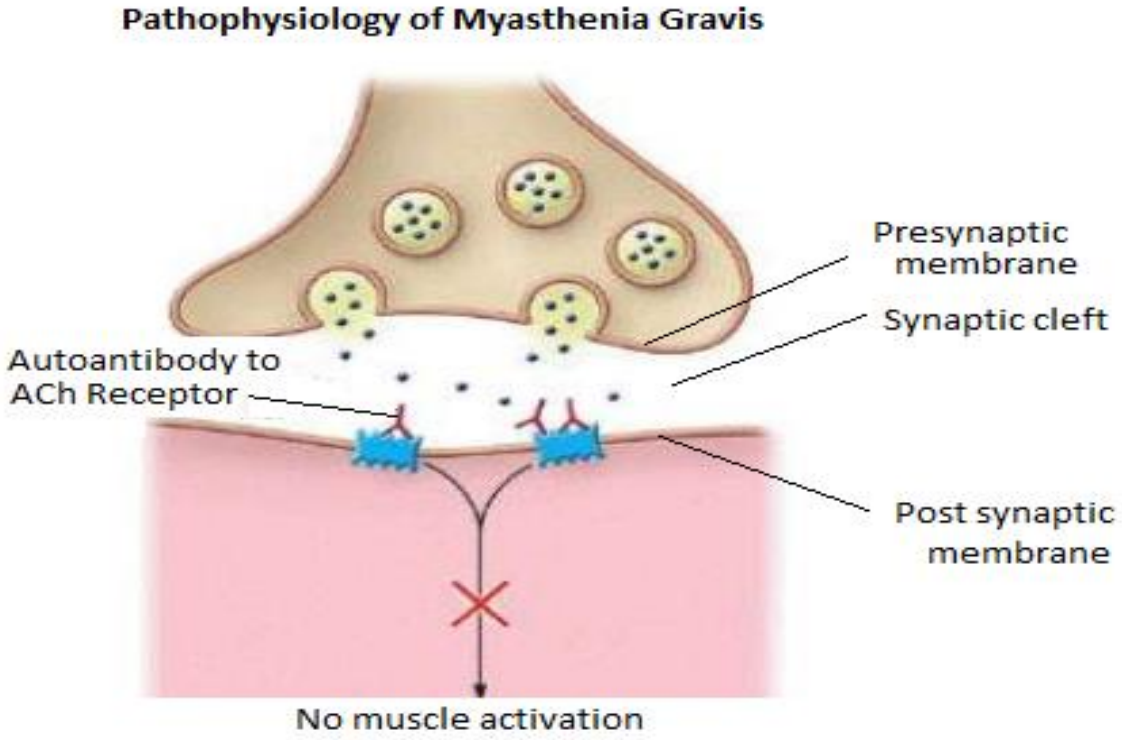
In the past, decompression surgery was used to relieve the pressure on the facial nerve by opening the bony passage that the nerve passes through. Today, decompression surgery isn't recommended. Facial nerve injury and permanent hearing loss are possible risks associated with this surgery.

Myasthenia gravis

Is a disease characterized by progressive muscular weakness on exertion, secondary to a disorder at the neuromuscular junction.

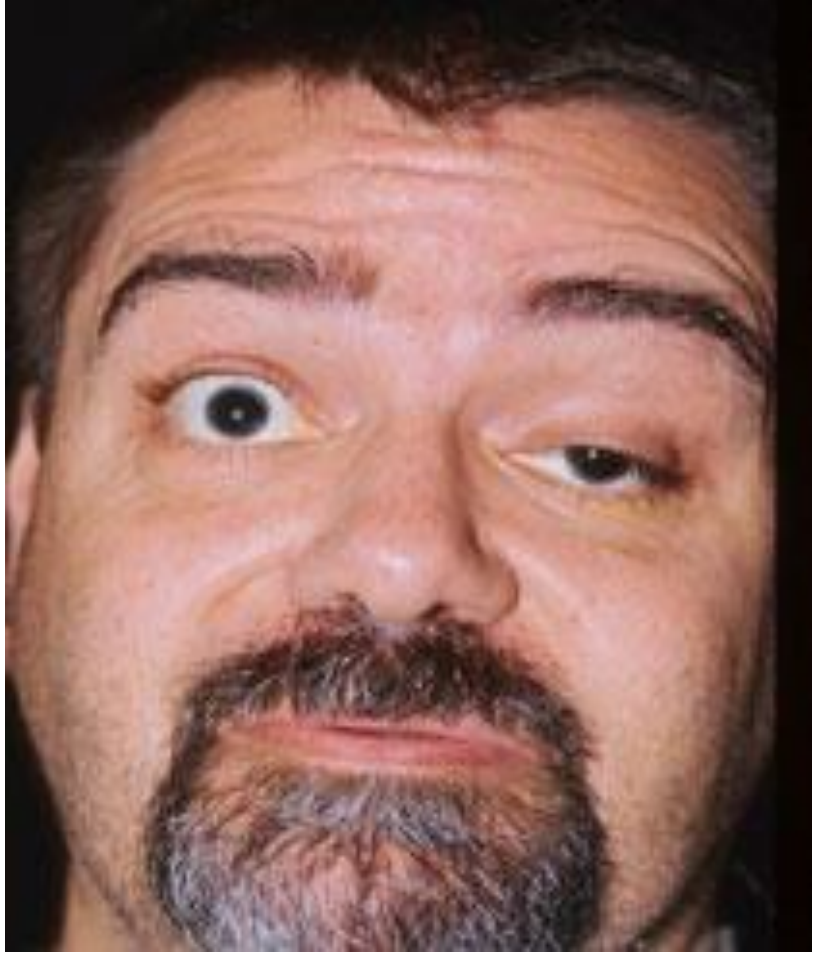
It is autoimmune disease, autoantibodies combine with and may destroy the acetylcholine receptor sites at the neuromuscular junction, preventing the transmission of nerve impulses to the muscle.

The initial signs of this disease commonly occur in areas innervated by the cranial nerves (frequently, the eye muscles).



General signs

1. Ptosis, diplopia
2. Difficulty in chewing or swallowing
3. Respiratory difficulties
4. Limb weakness



Oral and facial signs

1. The facial muscles of expression are involved
2. Tongue edema making eating difficult for patients
3. Difficulty in chewing; these patients will be unable to finish chewing a bolus of food because of the easy fatigability of the muscles

Treatment

1. Anticholinesterase drugs such as neostigmine and pyridostigmine bromide.
2. Thymectomy.
3. Long-term cortico-steroids and immunosuppressive drugs are necessary.

Dental management

1. A respiratory crisis may develop from the disease itself or from over medication.
2. Dental treatment should be performed in a hospital where endotracheal intubation.
3. The airway must be kept clear because aspiration may occur in patients whose swallowing muscles are involved.
4. Adequate suction and the use of a rubber dam are aids in these cases.
5. The dentist should avoid prescribing drugs that may affect the neuromuscular junction, such as: Narcotics, tranquilizers, and barbiturates.
6. Certain antibiotics, including tetracycline, streptomycin, sulfonamides, and clindamycin, may reduce neuromuscular activity and should be avoided.