Oral Medicine

Neuromuscular Disorders

Lecture 13

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Neuromuscular Disorders

Are diseases that affect both nerve and muscle tissue and represent a spectrum of nerve-related diseases and conditions that affect the body's voluntary muscles.

They cause weakening of muscles in the body because of interrupted communication between the nervous system and the muscles it controls.

Typically, these diseases can be managed to improve quality and length of life, but are incurable.

Classification of neuromuscular disorders:

- Cerebrovascular disease
- Multiple sclerosis
- Alzheimer's disease
- Parkinson disease
- Myasthenia gravis

Cerebrovascular disease:-

Cerebrovascular disease includes all disorders that cause damage to the blood vessels supplying the brain, leading to impaired cerebral circulation thereby producing neurologic damage.

Cerebrovascular accident (CVA) or Complete Stroke:

Is a sudden impairment in cerebral circulation resulting in death or a focal neurologic deficit lasting more than 24 hours.

Neurologic deficit: a term used to describe an acute neurologic injury resulting from a severe interruption in the flow of blood to the brain.

Neurologic events related to CVA include:

- **Transient ischemic attack (TIA):-**

A reversible, acute, short-duration, focal neurologic deficit (“mini stroke”) resulting from transient and localized cerebral ischemia (reversible within 24 hours).

- **Reversible ischemic neurologic defect (RIND):-**

A reversible, acute, focal neurologic deficit due to transient and localized cerebral ischemia but resulting in neurologic deficits that last more than 24 hours.

- **Stroke in evolution:** progressive worsening of stroke symptoms.

Causes of Cerebrovascular Accident (CVA) or Stroke :-

1. Atherosclerosis (85%) leading to cerebral ischemia and infarction result from ischemia due to atherosclerotic disease, thromboembolic events, and occlusion of cerebral blood vessels, with neurologic deficits related to the loss of neural function in tissues distal to the event.

2. Cerebral hemorrhage (15%) result from hemorrhagic events leading to infarction, most often related to hypertension, trauma, substance abuse, or aneurysmal rupture.

Clinical manifestations

The clinical manifestations of stroke vary depending on the size and location of the affected brain region.

<u>The most common signs and symptoms include:</u>

Sensory and motor deficits, changes (paresis) in extraocular muscles and eye movements, visual defects, sudden headache, altered mental status, dizziness, nausea, seizures, impaired speech or hearing, and neurocognitive deficits such as impaired memory, reasoning, and concentration.

Types of cerebrovascular diseases

Three major types of ischemic stroke syndromes have been described:

1. Small vessel (lacunar)
2. Large vessel (cerebral infarction)
3. Brainstem stroke

Lacunar strokes:-

- Result from obstruction of the small (<5 mm diameter) penetrating arterioles.
- Age and uncontrolled hypertension are the greatest predisposing factors.
- Symptoms usually include unilateral motor or sensory deficit without visual field changes or disturbances of consciousness or language.
- The prognosis for recovery from lacunar infarction is fair to good, with partial or complete resolution usually occurring over four to six weeks.

Cerebral infarction (large vessel):-

Is characterized by extensive downstream ischemia, usually due to a thromboembolic event along the distribution of the internal carotid artery and cerebral arteries.

Emboli often originate from the heart after acute myocardial infarction or in hyperdynamic conditions such as chronic atrial fibrillation.

Hypertension is an important risk factor in the development of thrombosis, particularly at the carotid bifurcation, and treatment of severe hypertension is essential for the prevention of stroke.

High level brain functions are affected, and the prognosis is poor.

Brainstem infarction :-

It is results from occlusion of small or large vessels supplying the brainstem, resulting in variable deficits ranging from motor and sensory deficits to death when respiratory centers are affected.

Diagnosis

In addition to a thorough neurologic and cardiovascular examination, anatomic and functional brain imaging is central to the diagnosis of stroke.
Brain magnetic resonance imaging (MRI) provides greater anatomic detail and sensitivity for detection of early infarction.

Treatment :-

The outcome of stroke and related TIAs and RIND is significantly affected by the timeliness of treatment.

Early intervention is critical to prevention, treatment and recovery.

TIAs and RIND are treated by reduction in hypertension (lifestyle changes such as diet, exercise, smoking cessation, and stress reduction; medical therapy for hypertension; and anticoagulant or antiplatelet medications).

Management of acute stroke includes medical therapy to reduce bleeding or thromboembolic occlusion, medical therapy to reduce brain edema and neurotoxicity/nerve injury, and surgical interventions (revascularization, hemorrhage control).

Once intracranial hemorrhage has been excluded as the source of acute cerebral ischemia, thrombolysis with intravenous tissue plasminogen activator (t-PA) can improve reperfusion, minimize infarction, and reduce disability.

After a completed stroke, treatment focuses on:

1. The prevention of further neurological damage, through the reduction of underlying risk factors
2. Rehabilitation procedures, including speech and physical therapy.
3. An intracranial hemorrhage should also be treated as a medical emergency of airway maintenance and requires the transfer of the patient to an intensive care unit with close monitoring.
4. The surgical treatment of a hemorrhaging aneurysm consists of closing off the blood vessels that supply the area and removing the abnormality.

Oral Health Considerations

1. Following stroke, patients may experience several oral problems, including masticatory and facial muscle paralysis, impaired or lost touch and taste sensation, diminished protective gag reflex, and dysphagia. These problems can lead to impairment of food intake, poor nutrition, and weight loss due to diminished taste satisfaction, chewing capacity, and swallowing; choking; and gagging.

2. Diminished motor function of masticatory and facial muscles may also reduce food clearance from the mouth and teeth with the presence of diminished dexterity of the arms or hands may adversely affect oral hygiene and increase the risk for caries and periodontal disease.

3. Prior history of TIA or stroke increases the risk of a future or second stroke, with the highest risk during the first 90 Days. With optimal medical monitoring and post-stroke care patients can safely undergo invasive dental treatment.

4. Use of antiplatelet and anticoagulant medications is common in patients with a history of stroke, TIA, and RIND may have a predisposition to excessive bleeding.

This includes oral aspirin, subcutaneous low-molecular- weight heparin and less commonly warfarin.

These medications taken in therapeutic dosages, and for warfarin with an international normalized ratio ≤ 3.5, rarely require dose modification before routine dental and minor oral surgical treatment.

It may be necessary to confer with the patient's physician to obtain current coagulation values (PT, INR) so as to ensure that the patient is stable for more invasive dental treatment.

5. Concomitant use of non-steroidal anti-inflammatory drugs (NSAIDs) may increase the risk for bleeding, and their long-term use may reduce the protective effect of aspirin.

6. Stress reduction and confidence building for the patient during dental visits are important behavioural goals to make the patient comfortable and minimize anxiety-related elevation in blood pressure. Pre-operative inhalation- N2O-O2 or oral anxiolytic medication can aid in reducing treatment related stress and anxiety.

7. Use of epinephrine-containing local anesthetics is not contraindicated, but they should be used wisely and follow guidelines recommended for patients with cardiovascular disease; epinephrine containing impression cord should not be used. Blood pressure should be monitored at every visit

8. Xerostomia is a common side effect of the medications used in the management of cerebrovascular disease. Patients can then be susceptible to a higher caries rate.

9. Careful history taking, checking of blood pressure prior to treatment, avoidance of lengthy appointments.

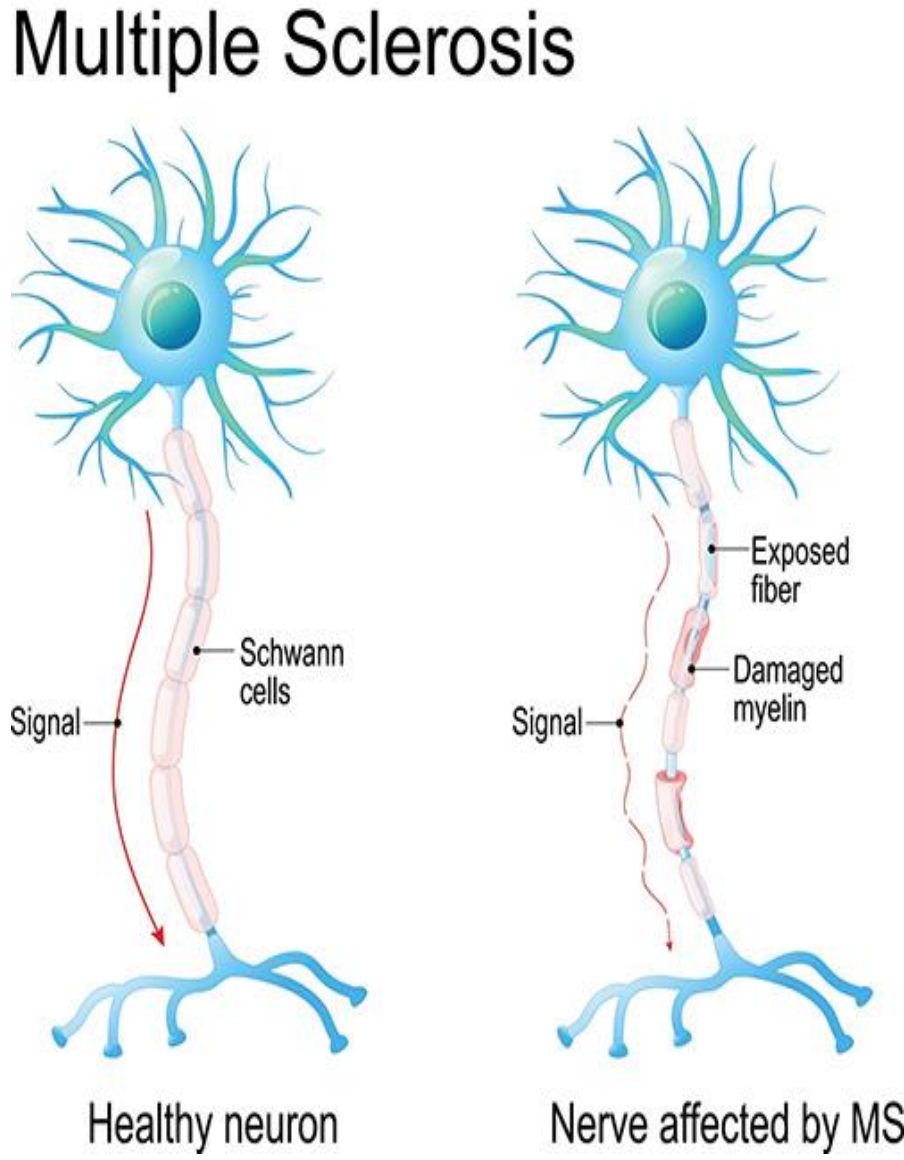
Multiple sclerosis (MS)

Is a chronic neurologic disease characterized by multiple areas of central nervous system (CNS) white matter inflammation, demyelination, and gliosis (scarring).

Myelin is critical for propagation of nerve impulses, and when it is destroyed in MS, slowing and/or complete block of impulse propagation is manifested by abnormal muscular and neurologic signs and symptoms, associated with the myelination of axons within the central nervous system.

The disease presents in the form of recurrent attacks and occurs more frequently among women.

The average age of onset is during the fourth decade of life, but MS may occur at any age.



Aetiology

1. An immunologic (autoimmune disease) basis is strongly suggested by the presence of activated T lymphocytes and autoantibodies to glycoproteins detected in MS lesions.

2. Environmental exposure in MS, and two common infectious agents to be implicated in the pathogenesis of this disease are Epstein–Barr virus and human herpesvirus. Other viruses that have been implicated in the pathogenesis of MS include measles, mumps, rubella, parainfluenza, vaccine and human T-lymphotropic virus.
   Increased antibody titers against measles virus, rubella virus, mumps virus, Epstein-Barr virus, herpes simplex viruses 1 and 2, and human herpes virus 6 (HHV-6) have been found in the cerebrospinal fluid and serum.

3. Genetic influences also appear to play a significant role in the development of MS.

Clinical Manifestations

The most common symptoms following an acute exacerbation include impairment of vision, muscular incoordination, and bladder dysfunction.

1. The clinical signs and symptoms of MS depend on the site of the demyelinating lesion of the CNS involved, and frequently affected areas include the optic chiasm, brainstem, cerebellum, and spinal cord.

2. More than 60% of individuals with MS have visual disturbances caused by demyelinating lesions of the second cranial nerve. The loss of vision usually occurs over a period of several days, with partial recovery within 1 month.

3. Other ophthalmic symptoms include “color blindness” and diplopia caused by involvement of the third, fourth, and sixth cranial nerves.

3. Uhthoff’s sign, found in MS, is characterized by rapid vision loss following a body temperature increase that is associated with strenuous exercise.

4. MS patients frequently complain of electric shock–like sensations that are evoked by neck flexion and radiate down the back and into the legs. This is referred to as Lhermitte’s symptom and is generally self limiting but may persist for years.

5. Weakness or paresthesia of the extremities, with an increase in the deep tendon reflexes, is another common early finding in cases of MS.

6. Bladder dysfunction, euphoria, ataxia, vertigo, and generalized incoordination.

7. The majorities of cases of MS are chronic and are characterized by exacerbations and remissions over a period of many years.

3. During acute episodes, severe neurologic involvement is evident. This slowly resolves, but some permanent neurologic involvement remains after each episode

Diagnosis

1. Clinically is based on the age of the patient, the presence of neurologic signs that cannot be explained by a single lesion, the progressive nature of the disease, and a history of exacerbations and remissions.

2. There are no definitive laboratory tests for MS, but demyelinating changes can be seen on (MRI) in more than 90% of cases. MRI demonstrates characteristic abnormalities of MS in >95% of patients. MS plaques are visible as hyperintense.

3. Evoked potentials measure CNS electrical potentials, and abnormalities are detected in up to 90% of patients with MS.

4. CSF is often analyzed in patients suspected of having MS, and positive findings include an increase in total protein and mononuclear white blood cells.

Treatment

1- High doses of **intravenous corticosteroids** may arrest the progress of MS; about 85% of patients with relapsing-remitting MS show objective signs of neurologic improvement during treatment with intravenous corticosteroids.
Glucocorticoids are used to manage both initial attacks and acute exacerbations of MS.
Intravenous methylprednisolone is typically administered at a dose between 500 and 1000 mg/d for three to five days to reduce the severity and length of an attacks

2- Long-term treatment with immunosuppressants may reduce the frequency of relapse in patients with MS. **Azathioprine** is probably the safest drug in this category and has reduced relapse to 70% of study patients in 3 years.
Administration of methotrexate appears to be the best therapy for slowing deterioration in patients with chronic progressive MS.

3- The use of interferon-γ-1b and -1a has shown promise; both have been shown to reduce clinical attacks and lesions

Oral Health Considerations

Individuals may present with signs and symptoms of MS.

1. Trigeminal neuralgia (TGN), which is characterized by electric shock–like pain, may be an initial manifestation of MS in up to 3% of cases.

MS-related TGN is similar to idiopathic TGN. Features of MS-related TGN include possible absence of trigger zones and continuous pain with lower intensity.
Medications often used to manage TGN are similar to those used for treatment of idiopathic TGN.

2. Patients with MS may also demonstrate neuropathy of the maxillary (V2) and mandibular branches (V3) of the trigeminal nerve, which may include burning, tingling, and/ or reduced sensation.

3. Neuropathy of the mental nerve can cause numbness of the lower lip and chin.

4. Myokymia may be seen in patients with MS and consists of rapid, flickering contractions of the facial musculature secondary to MS lesions affecting the facial nerve.

6. Facial weakness and paralysis may also be evident in MS patients.
7. Dysarthria that results in a scanning speech pattern is often seen in patients with MS.
8. TMJ disorder and headache.
9. Evaluate cranial nerve function, if cranial nerve abnormalities are detected, the individual should be referred to a neurologist for further evaluation.
10. It is recommended to avoid elective dental treatment in MS patients during acute exacerbations of the disease due to limited mobility and possible airway compromise.
11. Patients with significant dysfunction may require dental treatment in an operating room under general anesthesia due to the inability to tolerate treatment in an outpatient setting.
12. In addition, electric toothbrushes and oral hygiene products with larger handles may be necessary for completing oral hygiene in patients with significant motor impairment.
13. Be aware of possible interactions of these medications with those commonly used and prescribed in dentistry, as well as oral and systemic side effects of these agents.