SAQs Team

CASE 3

Case Scenario:
A 31-year-old **canadian woman** present with sudden onset of **right sided blindness** and with a strong family history of **autoimmune disease,** develops **ascending numbness** and **weakness in left feet** slightly asymmetrically, over a period of 2 weeks. She gradually develops **difficulty in walking.**

Regarding the case:

Q1 What are the differential diagnosis?
1- Multiple sclerosis (90 %.)
2- Myasthenia gravies (10%.)

Q2 What are the possible tests to diagnose the disease?
1- MRI.
2- Biopsy of CSF.

Q3 Mention the risk factors for developing this disease?
1- Female (young adult.)
2- Away from equator.
3- Family history.
4- Monozygotic twins.

Q5 What causes this disease?
Destruction of CNS myelin and oligodendrocytes.

Q6 Mention other symptoms that can develop?
1- Unilateral blurred vision.
2- Sexual dysfunction.

Q7 What is the management plan?
1-High dose of steroids
2- Interferon beta (slows progression of disease.)

Q8 Mention the gene related to this disease?
HLA-DR.

General questions

Q1 What are the functions of Myelin?

1-Insulator.
2-Increases the velocity of nerve impulses.

Q2 What are the functions of Oligodendrocytes and Microglia?

- Oligodendrocytes: Production of myelin sheath in CNS +optic nerve + insulation of nerve impulse.
- Microglia: phagocytosis.

Q3 What are the differences between CNS and PNS?

- Peripheral myelin is made by schwann cells, but CNS myelin is made by oligodendrocytes. Also each cell in PNS contribute to only one internode, while in the CNS many internodes come from a single oligodendrocyte.

Q4 What happens to myelin in both demyelinating and dysmyelinating diseases of the CNS?

Demyelinating diseases are acquired conditions which are characterized by preferential damage to normal myelin, while in the dysmyelinating diseases, myelin is not formed properly or has abnormal turnover kinetics.

Q5 What is the other general term for demyelinating diseases?

The other general term for dysmyelinating diseases is leukodystrophy.

Q6 What is shown in a patient who has Multiple Sclerosis?

The patient shows multiple episodes of relapses (symptoms) followed by episodes of remissions (recovery). Typically, the recovery is not complete.

Q7 What are the aim of most current treatments for Multiple Sclerosis?

They aim at decreasing the rate and severity of relapses rather than recovering the lost function.

Q8 What are the compositions of myelin?

- Lipids (80%): main component: Cerebrosides, other component:

sphingomyelin.
- Proteins (20%): for example, Myelin basic protein.

Q9 What is the name of the stain that we use it to detect Multiple Sclerosis?
Luxol fast blue/PAS.

Q10 What are the microscopic features that we can see in Multiple Sclerosis?
- Loss of myelin and variable loss of oligodendrocytes.
- Perivenous mononuclear inflammation (lymphocyte, plasma cell and macrophage).
- Relative preservation of axons.
- Reactive astrogliosis (sclerosis).
- Increased proportion of gamma globulin.
- Moderate pleocytosis.
- Oligoclonal bands representing antibodies.

Q11 What are the features that we see in early (acute) lesions of MS?
- Perivascular and parenchymal infiltration by inflammatory mononuclear cells, myelin breakdown and phagocytosis by macrophages.
- Astrogliosis is not profound and axons are relatively preserved.

Q12 What are the features that we see in chronic lesions of MS?
- There are few mononuclear cells, almost complete demyelination and severe astrogliosis.
- There can be oligodendrocyte loss and some secondary axonal loss in advanced cases.

Further important information:

The eye bulb:
There are three coats (tunics):
- Fibrous tunic: cornea and sclera.
- Vascular tunic: choroid, ciliary body and iris.

- Neural tunic: Retina.

There are four cranial nerves that supply the eye:
- Second cranial nerve: optic nerve.
- Third cranial nerve: oculomotor nerve.
- fourth cranial nerve: Trochlear nerve.
- sixth cranial nerve: Abducens nerve.

Optic nerve is a special sensory nerve and gives an important function which is it vision.

Oculomotor nerve supplies:
Motor (somatic fibres) to :
- Levator palpebrae superioris.
- Superior, medial and inferior rectus muscle.
- Inferior oblique.

Parasympathetic fibers to:
- Constrictor pupillae.
- Ciliary muscles.

Oculomotor is responsible for elevation of upper eyelid, turning the eye upward, downward and medially. Also responsible in constriction of the pupil and accommodation reflex of the eyes.

Trochlear nerve supply:
Motor to superior oblique (rotates the eyeball downwards and laterally)

Abducens nerve supply:
Motor to lateral rectus muscle (rotates the eyeball laterally (abduction))

Visual pathway:
-Optic nerve then goes to optic chiasma (where part of it decussates) then to optic tract after that to lateral geniculate body (nucleus) then they meet at optic radiation then to visual cortex.

If you have time , see this video
https://www.youtube.com/watch?v=M7O78LvrNSQ

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