

Answer to Medical Quiz -1

Answers:

1. Leonine facies.
2. Low voltage ECG.
3. Two-dimensional echocardiography left parasternal short-axis (C) and apical 4-chamber (D) views show significant left ventricular hypertrophy, granular appearance of myocardium and mild pericardial effusion. Image 1E represents bull's eye mapping of speckle-tracking echocardiography showing cherry-on-top appearance or apical sparing pattern.
4. Amyloidosis with cardiac involvement.
5. Apple-green birefringence under polarizing microscopy with Congo red staining.

Overview of amyloidosis with cardiac involvement

Amyloidosis is a heterogeneous group of disorders characterized by deposition of abnormal amyloid proteins in the body.¹ Amyloid deposits can build up in the heart, brain, kidneys, spleen and other parts of the body. A person may have amyloidosis in one or more organ. Mainly two types of amyloidosis affect the heart: AL amyloidosis and ATTR amyloidosis.² AL amyloidosis is closely related to plasma cell dyscrasia and is characterized by deposition of either kappa or lambda light chains in tissues.² On the other hand, ATTR amyloidosis occurs when the liver-derived protein transthyretin (TTR) misfolds and builds up in the organs and tissues.² These 2 types of cardiac amyloidosis produce similar findings in echocardiography, i.e., biventricular hypertrophy, biatrial enlargement, granular myocardium, systolic and diastolic dysfunction, and “cherry-on-top” appearance in bull's-eye mapping of strain

echocardiography.³ In fact, cherry-on-top pattern has 93% sensitivity and 82% specificity for cardiac amyloidosis.⁴ Significantly altered kappa-lambda light chain ratio in serum and monoclonal band in urine or serum in immunofixation electrophoresis are needed for the diagnosis of AL amyloidosis.³ For ATTR amyloidosis, preferential grade 2 or grade 3 radiotracer uptake by the myocardium in ^{99m}Tc-pyrophosphate scanning is suggestive.⁵ The diagnosis of amyloidosis is confirmed by histopathological examination of representative tissue showing apple-green birefringence under polarizing microscopy with Congo red staining.¹

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Answer to Medical Quiz -2

Answers:

- A. MRI of Cervical spine magnetic resonance imaging. (A1-B2) of sagittal slices demonstrate demyelinating lesions in the cervical spinal cord <2 segments). (A1, B1) T2-weighted images. (A2, B2) T1-weighted and postcontrast images.
- B. Primary progressive Multiple sclerosis
- C. I.V methylprednisolone followed by oral prednisolone
- D. Ocrelizumab and Ublituximab-xiiy

Overview:

Multiple sclerosis (MS) causes inflammatory demyelination and neurodegeneration in the brain and spinal cord. ^{1,2} MS is a multiphasic, chronic, relapsing demyelinating disease characterized by acute or subacute neurologic impairments. This condition primarily affects young to middle-aged females. Approximately 80% of MS patients acquire spinal cord lesions ³, which are more frequently symptomatic than brain lesions and can cause severe impairment such as ambulation, coordination, bladder and bowel function.

Spinal MS is frequently accompanied with concurrent brain lesions; nevertheless, up to 20% of patients with spinal lesions lack intracranial plaques. In contrast to the brain, both white and gray matter can be impacted in the spine. There is no substantial link between the size of the plaques and the level of clinical impairment. ³Spinal cord atrophy is mainly important to progressive forms of MS (primary and secondary progressive), when it is closely associated with physical disability. ⁴

The normal MRI involvement pattern is less than two cord segments, peripheral and ovoid appearance, and paracentral placement.⁵ MS is distinguished by the development of numerous demyelinating lesions in the brain and spinal cord that progress in time and space.⁶Typical features for MS in Spinal cord in Box:1.^{1,7}

Ocrelizumab is a humanised anti-CD20 monoclonal antibody used to treat multiple sclerosis (MS). The Food and Drug Administration (FDA) approved it in March 2017 for use in adults with RRMS and PPMS. Ocrelizumab is the sole disease-modifying treatment (DMT) approved for PPMS. ⁸ The FDA authorized Ublituximab-xiiy (Briumvi) in 2022. It is used to treat the relapsing-remitting and active secondary-progressive forms of MS. Ublituximab's method of

Box:1 Typical radiological features for MS in the spinal cord

- 1) There are multiple lesions in the spinal cord.
- 2) Typical spinal cord lesions in MS are relatively small and peripherally located.
- 3) They are most often found in the cervical cord and are usually less than 2 vertebral segments in length
- 4) In the cord there are some well-defined lesions, but also some ill-defined foggy lesions.
- 5) The transverse image shows the dorsal location and the typical triangular shape.
- 6) Continue with the contrast-enhanced image
- 7) Proton density weighted image (PDWI) is crucial for studying the spinal cord. On PDW-images the spinal cord has a uniformly low signal intensity (like CSF), which gives the MS lesions a good contrast against the surrounding CSF and normal cord tissue.

action involves the reduction of B cells by antibody-dependent cellular cytotoxicity, as B cells play an important part in the pathogenesis of MS. Ublituximab is the first anti-CD20 medication provided twice a year as one-hour infusions after the initial doses.⁹

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