

## Neurosarcoidosis: Review Questions

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### QUESTIONS

Choose the single best answer for each question.

- Neurosarcoidosis most often involves which of the following structures of the nervous system?**
  - Muscle
  - Meninges
  - Cerebellum
  - Cortex
  - Peripheral nerve
- A tissue sample positive for neurosarcoidosis exhibits nodules composed of all of the following cell types EXCEPT:**
  - Astrocytes
  - Lymphocytes
  - Endothelial cells
  - Giant cells
- The cranial nerve most frequently involved in neurosarcoidosis is:**
  - VII
  - III
  - II
  - IV
  - I
- All of the following are neurosarcoid-associated syndromes EXCEPT:**
  - Aseptic meningitis
  - Diffuse encephalopathy with vasculopathy
  - Amyloidosis
  - Seizures
  - Hypothalamic syndrome
- All of the following laboratory data are supportive of sarcoidosis EXCEPT:**
  - Increased serum globin levels
  - Increased serum calcium levels
  - Increased serum/cerebrospinal fluid angiotensin converting enzyme (ACE) levels
  - Positive skin-tuberculin reaction
  - Increased glycol-precipitable immunoglobulin levels
- Which of the following choices best describes how the diagnosis of neurosarcoidosis is made?**
  - Clinically
  - By a history of cranial neuropathy and an elevated ACE level in serum
  - By confirmatory biopsy of a nodule and a history of neurologic signs or symptoms
  - Only at autopsy
- The differential diagnosis of neurosarcoidosis includes all of the following EXCEPT:**
  - Hodgkin's lymphoma
  - Non-Hodgkin's lymphoma
  - Syphilis
  - Multiple sclerosis

*(turn page for answers)*

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**EXPLANATION OF ANSWERS**

1. **(B) Meninges.** Although sarcoidosis may involve any part of the nervous system, it most often involves the meninges. Such involvement is often manifested by aseptic meningitis and/or peripheral neuropathy, particularly cranial neuropathy.
2. **(A) Astrocytes.** Astrocytes are not found in sarcoid nodules. Biopsies of sarcoid nodules are composed of lymphocytes, endothelial cells, and giant cells.
3. **(A) VII.** The cranial nerve most often involved in neurosarcoidosis is cranial nerve VII. Other cranial nerves may also be involved, in the following order of frequency: VIII, II, III, IV/VI, and I. It should be noted that facial paralysis usually occurs in association with uveitis and parotitis.
4. **(C) Amyloidosis.** Amyloidosis is not associated with neurosarcoidosis. Neurologic syndromes associated with neurosarcoidosis include aseptic meningitis, diffuse encephalopathy with vasculopathy, and seizures. The presence of seizures worsens the prognosis. In addition, a hypothalamic syndrome has been described. This includes clinical manifestations such as obesity, chronic hypernatremia, hypovolemia, and frank diabetes insipidus. Any peripheral nerve may be involved in sarcoidosis, with or without other cranial or peripheral nerve involvement.
5. **(D) Positive skin-tuberculin reaction.** Patients with sarcoidosis often exhibit cutaneous anergy. Serum globulin is increased in sarcoidosis, although it is a non-specific marker of disease. Increases in ACE levels have been seen in some patients with active disease. Circulating immune complexes are elevated (especially glycol-precipitable immunoglobins), as is serum calcium.
6. **(C) By confirmatory biopsy of a nodule and a history of neurologic signs or symptoms.** The diagnosis of neurosarcoidosis is made by observing an appropriate clinical history with neurologic signs or symptoms, accompanied by findings on the biopsy of a nodule or affected tissue consistent with sarcoidosis.
7. **(B) Non-Hodgkin's lymphoma.** Hodgkin's lymphoma, syphilis, and multiple sclerosis are all part of the differential diagnosis of neurosarcoidosis.

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**NEUROLOGY**

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