

Scleroderma

MCQs

- typical case scenario of scleroderma with shortness of breath , what is the most likely diagnosis?

Pulmonary hypertension

- In which of the following is more likely to have positive anti-centromere antibody.

a. CREST syndrome

b. Diffuse Scleroderma.

c. Mixed Connective Tissue Disease

d. SLE (systemic lupus erythematosus)

e. Vasculitis.

- Wrong regarding scleroderma :

there is vegetation on the heart valves

- Wrong about scleroderma:

A. Pulmonary fibrosis

B. Encephalopathy

Answer: B.

Pulmonary involvement — Pulmonary involvement is seen in more than 70 percent of patients with SSc. The two principal clinical manifestations are interstitial lung disease (also called fibrosing alveolitis or pulmonary fibrosis) and pulmonary vascular disease, leading to pulmonary arterial hypertension ([table 5](#)). These issues are discussed in detail separately but will be briefly reviewed here. (See "[Clinical manifestations of systemic sclerosis \(scleroderma\) lung disease](#)".)

Neuromuscular involvement — Neuromuscular involvement in SSc is discussed in more detail elsewhere. (See "[Neuromuscular manifestations of systemic sclerosis \(scleroderma\)](#)".) The following is a brief summary of the types of neurologic and muscle disorders that have been noted in case reports and series:

- Cranial, entrapment, peripheral, cutaneous, autonomic neuropathies
- Myopathy and inflammatory myositis
- Central nervous system involvement, including headache, seizures, stroke, vascular disease, radiculopathy, and myelopathy

- Which antibody is rather specific for diffuse scleroderma?
 - a. Anticentromere AB
 - b. Anti-myeloperoxidase AB (p-ANCA)
 - c. Anti-Jol AB
 - d. Antimitochondrial AB
 - e. Anti-Scl70**

Answer: E [Diagnosis is by clinical suspicion, presence of autoantibodies (specifically anti-centromere and anti-scl70/anti-topoisomerase antibodies) and occasionally by biopsy. Of the antibodies, 90% have a detectable anti-nuclear antibody. Anti-centromere antibody is more common in the limited form (80-90%) than in the diffuse form (10%), and anti-scl70 is more common in the diffuse form (30-40%) and in African-American patients (who are more susceptible to the systemic form).[19]

In 1980 the American College of Rheumatology agreed upon diagnostic criteria for scleroderma.[20]

- All the following are poor prognostic signs in scleroderma except.
 - a- old age of onset.
 - b- limited skin involvement.**
 - c- high ESR
 - d- renal involvement
 - e- pulmonary hypertension
- Associated with Raynaud (systemic sclerosis)

Mini-OSCE

Q3 - A 42 year old female with finger pain upon cold exposure, If this patient's blood pressure is 150/90, what drug would you choose?

- a. Beta blockers
- b. ACE inhibitors**
- c. Thiazides
- d. Vasodilators
- e. Calcium channel blockers

