

Test your knowledge

Scleroderma is a disabling multisystem autoimmune disease. In the past, malignant hypertension meant that scleroderma was a lethal condition, but that has changed with the introduction of ACE inhibitors, and most deaths associated with scleroderma are now due to pulmonary fibrosis and hypertension. Would you recognise scleroderma in your practice?

The multiple choice questions in this quiz may have more than one answer.

- What is the typical skin lesion in scleroderma?
 - pitting oedema
 - a butterfly rash over the face
 - vasculitic ulcers on the legs
 - thickened, hard skin
 - associated psoriasis
- What other abnormalities are seen in the hands of people with scleroderma?
 - telangiectasia
 - calcium deposits in the pulp of the fingertips
 - flexion contractures caused by thickened skin
 - destructive arthritis of the small joints
 - sclerodactylia
- Raynaud's phenomenon is very commonly associated with scleroderma (see Figure 1). Which of the following are true?
 - Raynaud's phenomenon is a change in the colour of the digits that is precipitated by cold or occasionally by vibration
 - Raynaud's phenomenon is a change in the colour of the digits from white, through blue, to red
 - Raynaud's phenomenon is a change in the colour of the digits from red, through blue, to white
 - Raynaud's syndrome is caused by vasoconstriction
 - Raynaud's syndrome is caused by a hyperviscosity syndrome
- What might help to reduce the frequency of attacks of Raynaud's phenomenon?
 - moving to a cold climate
 - always wearing gloves in cold weather (one pair or possibly two)
 - avoiding 'hot' showers and using tepid water instead
 - not eating chillis or drinking alcohol
 - moving to a warmer climate
- Oesophageal symptoms are common in scleroderma. What are the causes?
 - oesophageal webs causing dysphagia
 - ulcerating reflux oesophagitis causing heartburn
 - dysphagia caused by a motility disorder of the oesophagus
 - achalasia
 - squamous cell carcinoma of the oesophagus
- Endoscopy is indicated in patients with scleroderma who complain of retrosternal burning. What could be used to treat the changes seen in Figure 2?
 - fluconazole
 - an antacid
 - ranitidine, 150 mg twice daily
 - domperidone, 10 mg twice daily
 - omeprazole, 20 mg twice daily



Figure 1.

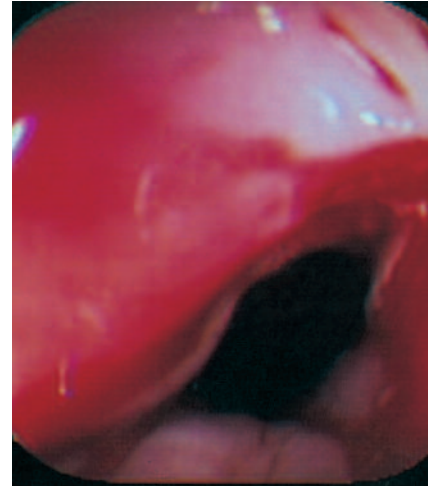


Figure 2.

PHOTOGRAPHS COURTESY OF DR KATIE ELLARD, SYDNEY, NSW

Reference

- Moxley G. Scleroderma and related diseases. In: Rubinstein E, ed-in-chief. Scientific American medicine. New York: Scientific American, 1996; 15(v).

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1. d

The classic abnormality in scleroderma is thickened, hard skin. The distribution varies from patient to patient.

2. a, b, c, e

Sclerodactylia is a deformity caused by the thickened, nonpitting swelling over the digits of the skin of the fingers that leads to contractures. Calcium deposits on the tips of the fingers, ulcers and telangiectasia are also common. An arthritis resulting in joint destruction is much more typical of psoriasis or (occasionally) rheumatoid arthritis.

3. a, b, d

Raynaud's syndrome is a change in colour in the digits precipitated by cold or occasionally by vibration. It is caused by vasoconstriction followed by a reactive hyperaemia, and can result in ulceration of the fingertips and gangrene.

4. b, e

Raynaud's syndrome is precipitated by cold temperatures – therefore, cold climates make it worse, and it is worth advising the patient to invest in a good pair of fleecy gloves. Double gloving is advised in very cold temperatures. Some patients respond to vasodilators.

5. b, c

The lower oesophageal sphincter does not work adequately in scleroderma, allowing marked acid reflux. Abnormalities of smooth muscle lead to poor peristalsis in the oesophagus and give rise to dysphagia.

6. e

Figure 2 shows ulcerating reflux oesophagitis, which is common in scleroderma. Fluconazole is used to treat oesophageal candidiasis, which is not more common in people with scleroderma than in the general population, unless they are on medication that suppresses the immune system or on long term antibiotics. An antacid or H₂ antagonist is not sufficient in ulcerating reflux oesophagitis associated with scleroderma. A proton pump inhibitor is required, often twice a day. Domperidone is useful for motility disorders that may continue to cause dysphagia or reflux symptoms after adequate acid suppression.

7. d

Sjögren's syndrome is seen in approximately one-third of people with scleroderma. The excessively dry mouth can exacerbate swallowing difficulties.

8. c

Approximately 95% of people with scleroderma are positive for antinuclear antibodies, although the test is not specific for this condition. A positive endomysial antibody is indicative of coeliac disease; antimitochondrial antibody is associated with primary biliary cirrhosis. Rheumatoid factor is associated with rheumatoid arthritis, and smooth muscle antibody is associated with autoimmune chronic active hepatitis.