Scleroderma

Scleroderma
• Connective tissue disorder: fibrosis
• Arteriolar abnormalities: Raynaud’s
• Autoantibodies present
• Spectrum of severity
• Hand changes: sclerodactyly
• Lung, heart, gut and renal involvement: systemic sclerosis

Hands: inspect
• Tight, thickened, tethered skin with shiny appearance: “waxy”
• Loss of skin creases, especially over knuckles
• Colour changes of Raynaud’s
• Tapering fingers with pulp atrophy and small finger pulp scars or “pits”; skin ulcers
• Telangiectasia
• Synovitis of small joints and tendons, often with fixed flexion: positive prayer sign

Hands: palpation
• Temperature: cold
• Telangiectasia: blanch with pressure
• Reduction in normal skin movement: tethering
• Subcutaneous calcification
• Reduced hand function- grip strength may be impaired and loss of finger extension

Arms
• Check to see if scleroderma extends onto arms with reduced skin movement
• Check for subcutaneous calcification (typically forearms)
• Ask for or measure the blood pressure (underlying renal disease)

Face: inspect
• Radial furrows around mouth
• Reduced mouth opening: microstomia
• Tight skin over forehead and narrowed bridge of nose
• Multiple telangiectasia over cheeks
Face: palpation
• Reduced skin movement over forehead, bridge of nose and around mouth
• Telangiectasia- blanching

Investigations
• Raised ESR and CRP; FBC may show normochromic normocytic anaemia if renal failure present
• Renal function: check urine, plus serum urea, creatinine and electrolytes
• LFT as rare association with primary biliary cirrhosis
• Barium swallow for dilated atonic oesophagus
• CXR and lung function tests: pulmonary fibrosis
• ECG (myocardial fibrosis and changes due to hypertension) +/- echo
• Autoantibodies: anti-nuclear factor, typically with speckled appearance; anti-centromere and anti-nucleolar subtypes are common