Rare cause of red eye in scleroderma

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DESCRIPTION

A 37-year-old woman with Raynaud's phenomenon in all fingers and toes for 7 years presented with painful redness of both eyes for 2 weeks. On examination, she had digital ulcers (figure 1), skin tightening with microstomia (modified Rodnan's skin score was 15/51), bilateral vision was 6/6, with normal intraocular pressure. However, the anterior sclera showed focal inflammation on the nasal side with dilated scleral and episcleral vessels, findings suggestive of nodular scleritis bilaterally, with normal posterior segments and retinal vasculature (figure 2).

On investigations, she had elevated erythrocyte sedimentation rate and C-reactive protein, antinuclear antibody was positive by enzyme immuno assay, Scl 70 was equivocal and anticentromere was negative. She was diagnosed as diffuse cutaneous systemic sclerosis with bilateral nodular scleritis . High-resolution CT thorax showed non-specific interstitial pneumonitis pattern of interstitial lung disease. Pulmonary function testing revealed a restrictive pattern of lung disease with a forced vital capacity of 59%. Her rheumatoid factor, antiproteinase 3/antimyeloperoxidase, hepatitis B surface antigen, antihepatitis C, venereal disease research laboratory test, anticitrullinated peptide antibodies and Mantoux test were negative. There was no evidence of pulmonary artery hypertension on ECG or two-dimensional echocardiography. She was started on non-steroidal anti-inflammatory drugs, oral steroids 1 mg/kg with monthly pulses of injection cyclophosphamide (0.5 g/m^2 of body surface area). On follow-up 4 weeks later, she had marked reduction in inflammation with healing scleritis bilaterally.

Scleral involvement has been described with systemic autoimmune diseases, notably rheumatoid arthritis, granulomatosis with polyangiitis, sarcoidosis, systemic lupus erythematosus or spondyloarthritis.[1] Ocular involvement in scleroderma has been studied in few series[2 3] where the most common findings were eyelid stiffness, eyelid telangiectasia, keratoconjunctivitis sicca, astigmatism, cataract, glaucoma and retinal vascular abnormalities. Orbital inflammation and necrotising scleritis have been reported with limited scleroderma.[4 5] This index case highlights that scleritis, though extremely rare, can also be seen in diffuse scleroderma and requires early intervention.

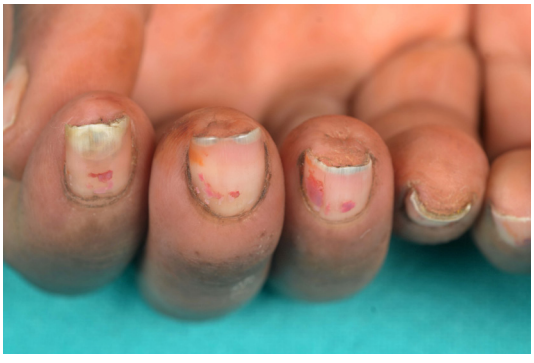

Figure 1 Image showing digital ulcerations at the fingertips.

Figure 2 Image showing focal inflammation at the nasal side of right sclera suggestive of nodular scleritis.

Learning points

- Scleritis has been described with various autoimmune diseases, but is a very rare association with scleroderma.
- Early identification and treatment can prevent complications/sequelae due to scleritis.

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