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Scleroderma and Sjögren's syndrome

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Scleroderma is a rare condition, occurring most commonly in females, the onset of which is usually heralded by Raynaud's phenomenon, cutaneous sclerosis, or arthritic symptoms. General constitutional disturbances such as lassitude, anorexia, and weight loss are common. The course of the disease may be prolonged, or death may supervene within a year of onset. No part of the body appears to be immune from the disease process.

This communication reports a case of Sjögren's syndrome in association with scleroderma and presents the ocular findings in nine other cases of scleroderma.

Case report

A 62-year-old female (Case 10, see Table) presented complaining of a gritty sensation in both eyes of 2 years' duration. For 20 years she had suffered from Raynaud's phenomenon, terminal ulceration of the digits, and arthritis of the hands, and over the previous 10 years her skin had become smooth, shiny, and taut, the face and upper limbs being particularly involved. For 2 years she had also suffered from xerostoma and dysphagia.

Table *Ocular findings in ten cases of scleroderma*

Case No.	*Age (yrs)*	*Duration of disease (yrs)*	*Palpebral apertures (mm.)*	*Schirmer's test*		*Corneal stain*	*Cataract*
				Right	*Left*		
1	67	20	7 × 25	15	12	—	Yes
2	55	20	9 × 28	5	8	—	Yes
3	52	6	8 × 24	4	9	Yes	Yes
4	51	10	9 × 27	15	12	—	Yes
5	47	13	8 × 25	0	2	Yes	—
6	76	12	10 × 28	15	12	—	Yes
7	57	5	10 × 27	10	5	—	—
8	70	5	8 × 27	15	16	—	—
9	57	6	9 × 28	14	15	—	—
10	62	20	8 × 22	8	3	Yes	Yes

Examination General emaciation was present. The facial skin was waxy and inelastic and did not move over the facial skeleton. The nose was pinched and the mouth small and puckered. Occasional telangiectases were present. The fingers were fixed in a flexion deformity with the appearance of rheumatoid arthritis and terminal ulcers were present on three digits of the left hand.

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Ocular examination The skin of the lids was inelastic and the upper lid folds almost indiscernible. The palpebral apertures were much reduced in size, measuring 8 × 22 mm. both right and left.

The corrected visual acuity was 6/24 in the right eye and 6/9 in the left. Slit-lamp examination revealed a thin slow-moving precorneal film. Filamentous keratopathy with extensive rose Bengal and fluorescein staining was present. There were moderate cortical lens opacities and the fundi were normal. Schirmer's test showed diminished tear secretion (right 8 mm. and left 3 mm.) after 5 minutes.

Treatment The keratopathy was controlled within 2 weeks by steroid and methylcellulose drops, and she was subsequently treated with methylcellulose drops only.

Progress Some 3 months later she presented with almost total de-epithelialization of the right cornea and extensive staining of the left cornea. The conjunctivae were remarkably white. Treatment in hospital with local atropine and antibiotics, the eyes being padded, rapidly settled this episode.

After a further 3 months she presented with extensive staining of the right cornea and a corneal abscess. No pathogenic organisms were isolated from conjunctival cultures. Despite intensive antibiotic treatment, local and systemic, the cornea perforated and the eye was, therefore, eviscerated. Histological examination of the cornea revealed no evidence of viral or fungal infection.

The sclerodermatous process progressed rapidly in the relatively unsupported right eyelids and she subsequently had difficulty in inserting her artificial eye. A lateral canthoplasty has helped in this respect.

Result At the present time the palpebral apertures are reduced to 8 × 22 mm. right and 8 × 20 mm. left. Filamentary keratopathy and rose Bengal staining of the left cornea persist. The patient admits that she uses the methylcellulose drops only sporadically.

Nine further female patients were examined whose ages ranged from 47 to 76 years and in whom the scleroderma had been manifest for from 5 to 20 years. The relevant ocular findings are presented in the Table.

Discussion

The ocular manifestations of scleroderma have excited little attention in the literature. Vail (1952) mentioned lid thickening and pigmentation and cataract formation. In the present series of cases four patients demonstrated a degree of blepharophimosis which was strikingly symmetrical. The normal adult palpebral apertures measure 10 × 28 mm. (Fox, 1966). Pigmentation and telangiectases involving the skin of the lids were present in two cases; in the remainder the lids were not involved.

Presenile cataracts of sufficient degree to reduce visual acuity were present in three cases (2, 3, and 4). The cataracts noted in Cases 1, 6, and 10 are thought to be related not to scleroderma but to the patients' ages. However, in a series of fourteen cases of scleroderma, Stucchi and Geiser (1967) detected no significant lens opacification and concluded that cataract was not part of the clinical picture of scleroderma.

There are few reports of the development of keratoconjunctivitis sicca in scleroderma. Discussing keratoconjunctivitis and the collagen diseases, Ramage and Kinnear (1958) mentioned only one case of scleroderma. In a review of 248 cases of keratoconjunctivitis sicca only two were noted to have scleroderma (Stoltze, Hanlon, Pease, and Henderson, 1960). Two cases of scleroderma in which reduced lacrimal secretion, measured by Schirmer's test, was present were described by Shearn (1960). The diagnosis of scleroderma in the case reported by Harrington and Dewar (1951) in association with Sjögren's syndrome has been questioned (Shearn, 1960). Diminished lacrimal secretion, measured by Schirmer's test, was found in eleven of fourteen patients by Stucchi and Geiser (1967), although Sjögren's syndrome was not seen in this series.

In the present series of cases one patient was found to have Sjögren's syndrome, and there was evidence of reduced lacrimal secretion in four others. Two of the latter showed corneal staining with rose Bengal but no filamentary keratopathy. In each case lacrimal secretion was measured by Schirmer's test over a period of 5 minutes under identical conditions and 12 mm. was accepted as the lower limit of normal.

Retinopathy is rarely associated with scleroderma. Agaston (1953), Pollack and Becker (1962), and Klien (1965) each saw one case. Hypertension or renal disease was present in all three, and there is no reason therefore to implicate scleroderma as the cause of the retinopathy. In the present series, as in that of Stucchi and Geiser (1967), no case of retinopathy was observed. Nor were colloid bodies seen of the type described by Hartmann, Collin, and Vergne (1948).

Summary

A case of scleroderma with Sjögren's syndrome is described and the ocular findings in nine other patients with scleroderma are tabulated. Lacrimal hyposecretion was present in five patients. It is believed that there is a definite association between scleroderma and lacrimal hyposecretion with resultant keratoconjunctivitis sicca.

Presenile cataract appears to constitute part of the clinical picture of scleroderma.

When the lids are involved by the sclerodermatous process a degree of symmetrical blepharophimosis results.

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