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Ophthalmic manifestations of progressive systemic sclerosis

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The ocular manifestations of progressive systemic sclerosis (PSS) have been described by Manschot (1965), Nelken and Michaelson (1961), Pollack and Becker (1962), and Stucchi and Geiser (1967). These reports were mainly limited to isolated cases, although Stucchi and Geiser (1967) studied a series of fourteen patients—thirteen with generalized scleroderma and one in whom the scleroderma was circumscribed.

Material and methods

A series of 23 patients (nineteen females and four males) suffering from PSS, all of whom were currently being followed up in the Department of Dermatology at the Leeds General Infimary, was studied. Cases of morphea and Werner's syndrome were excluded. For the purposes of this report, systemic sclerosis is defined as "a disorder in which the features of Raynaud's phenomenon are followed by changes of an atrophic and sclerotic nature, associated with widespread vasculitis, in the skin and internal organs" (Rowell, 1968).

A control group of 23 subjects not suffering from PSS—and carefully matched for age and sex—was selected principally from among members of the hospital staff. The age distribution of patients and controls is shown in Table I.

Table I *Age groups and mean age of patients and controls*

Age group (yrs)	0–10	11–20	21–30	31–40	41–50	51–60	61–70	71–80	*Total cases*	*Mean age (yrs)*
Patients			2	3	8	8	0	2	23	48
Controls			2	3	8	8	0	2	23	49·5

The ophthalmic examination included Schirmer's test, slit-lamp microscopy, fluorescein and rose bengal staining, applanation tonometry, and ophthalmoscopy under a mydriatic. Blood pressure and urinary examinations were also carried out.

Schirmer's test was performed using a strip of filter paper 35 mm. long and 5 mm. wide. One end of the strip was placed into the lower conjunctival fornix and the remainder folded at right angles over the lid margin, and the wetting of the strip was read after 5 minutes. The test was regarded as positive when there was less than 5 mm. wetting; between 5 and 15 mm. was regarded as suspicious, and over 15 mm. as normal.

Rose bengal 1 per cent. solution was instilled into the conjunctival sac of each eye, and immediately followed by irrigation with normal saline. The patients were then examined by a Haag Streit-900 slit lamp for staining patterns and filamentary keratitis (this test was carried out before the Schirmer's test to avoid staining due to contact with the filter paper).

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Findings

LESIONS OF THE EYELIDS

Telangiectases of the upper and lower lids of both eyes were present in four patients. This was part of a more generalized distribution of similar lesions on the face. Three further patients had facial telangiectases without lid involvement.

Tightness of the lids, both upper and lower, was present in fifteen cases. This varied from a slight immobility and loss of elasticity to the more severe cases where the lower lids could not be everted.

REDUCED TEAR SECRETION

Eleven (48 per cent.) of the present group of patients had diminished tear secretion.

Keratoconjunctivitis sicca was present in seven patients and in addition two of these had xerostomia and rheumatoid arthritis, *i.e.* Sjögren's syndrome.

These seven patients all had symptoms due to reduced tear secretion. They also had rose bengal staining of the conjunctiva. Schirmer's test was positive.

Two patients had involvement of one eye only.

Hyposecretion of tears was found in four symptomless patients as an isolated finding.

CONJUNCTIVA, CORNEA, AND SCLERA

Shallow conjunctival fornices were present in five cases. One patient had a mild conjunctival oedema and three a chronic non-specific conjunctivitis. There was no evidence in these cases to support a viral or bacterial aetiology. A total of fifteen patients (65 per cent.) had rose bengal staining of the conjunctiva and in eight of these it was present as an isolated phenomenon. One case of keratoconjunctivitis sicca had a filamentary keratitis which was limited to the lower third of the cornea. No other patient had any corneal abnormality. Finally, one patient was suffering from a recurrent nodular episcleritis.

LENS

Very early lens opacities were present in eighteen patients. The majority were early senile lens opacities but some were developmental in origin. Impairment of vision was not a prominent feature, although one woman aged 59 years had a mature cataract in the right eye. A history of trauma was not elicited, and ocular transillumination and orbital *x* rays were normal.

FUNDUS

A small superficial retinal haemorrhage was observed in one 48-year-old woman with normal vision. There was no history of trauma or evidence of diabetes, hypertension, or arteriosclerosis. Two patients had colloid bodies at the posterior pole and one had a patch of old choroiditis.

Control group

Examination of the control group of 23 subjects, matched for age and sex but not suffering from PSS, produced interesting results.

Early lens opacities were present in seventeen, but in fifteen of them there was no visual impairment. Two, aged 58 and 72 years, had more advanced senile lens opacities, causing slight reduction in vision.

Reduced tear secretion was found in one case.

Rose bengal staining of the conjunctiva was found as an isolated sign in nine cases; the cornea was not involved.

Two control cases had bilateral colloid bodies at the posterior pole; there were no other abnormalities of the fundus.

Statistical analysis

Table II, which compares the incidence of eye disease in the patients with PSS with that in the control subjects matched for age and sex, provided interesting material for statistical analysis. For example, lens opacities of some form or another were found in eighteen patients with PSS and in seventeen of the control group. The presence of lens opacities in patients with PSS can thus probably be attributed to their age rather than to their systemic disease, but this is not the case with the other ophthalmic manifestations. Fifteen out of 23 patients with PSS had lid tightness, eleven had diminished tear secretion, seven had keratoconjunctivitis sicca, five had shallow conjunctival fornices, and four had telangiectasis of the lids. Comparable findings were not demonstrated in the control group and it would therefore seem that these ophthalmic features are manifestations of PSS. The significance of the difference is shown in Table II.

Table II *Ophthalmic involvement in patients and controls*

Lesions	*Number of cases*		χ^2	*n*	*P<*
	Progressive systemic sclerosis (23)	*Controls* (23)			
Lens opacities	18	17	0·03	1	>0·90
Lid tightness	15	0	15·00	1	<0·001
Rose bengal staining of conjunctiva	15	9	1·50	1	0·1>P >0·05
Hyposecretion of tears	11	1	8·33	1	<0·01
Keratoconjunctivitis sicca	7	0	*	–	<0·01
Shallow fornices	5	0	*	–	<0·03
Telangiectases	4	0	*	–	0·1>P >0·05
Sjögren's syndrome	2	0	*	–	>0·2

*χ^2 inapplicable because of small numbers P estimated by absolute method

The presence of other manifestations of eye disease, such as keratoconjunctivitis sicca in association with xerostomia and arthritis in patients with PSS is less easy to analyse. Certainly this condition is rare in the general population at large, and it is of interest that it should have been found in two of this small group of patients, although the numbers are too small for conclusions to be drawn.

Discussion

The literature abounds with a multitude of confusing reports which suggest that there may be a link between progressive systemic sclerosis and eye disease (Hartman, Collin, and Vergne, 1948; Agatston, 1953; Leinwand, Duryee, and Richter, 1954; Rossier and Hegglin Volkmann, 1954; Ramage and Kinnear, 1956; Štáva, 1958; Munro, 1959; Salgado

Gomez, 1960; Shearn, 1960; Stoltze, Hanlon, Pease, and Henderson, 1960; Nelken and Michaelson, 1961; Tuffanelli and Winkelmann, 1961; Pollack and Becker, 1962; Block and Bunim, 1963; Rodnan, 1963; Vanselow, Dodson, Angell, and Duff, 1963; Goder, 1964; Manschot, 1965; Sanders, 1966; Stucchi and Geiser, 1967). Unfortunately many of these reports contradict one another, drawing mutally exclusive conclusions, and many of them are based upon isolated case histories.

However, Stucchi and Geiser (1967) reported the ophthalmic manifestations in fourteen patients (ten females and four males) suffering from generalized progressive systemic sclerosis. Hyposecretion of tears was present in eleven cases, but keratoconjunctivitis sicca and/or Sjögren's syndrome were not found. These Swiss workers found cataracts in three patients, but they concluded that two were probably senile cataracts, and that in the third case the lens opacity was probably secondary to uveitis and glaucoma. They also reported a progressive reduction in the conjunctival fornix.

The present controlled study demonstrates that specific ophthalmic lesions do occur in association with PSS, the most common being tightness of the lids, diminution in the tear secretion, keratoconjunctivitis sicca, telangiectasia, and shallow fornices. Occasionally xerostomia and arthritis accompanied keratoconjunctivitis sicca.

KERATOCONJUNCTIVITIS SICCA

Duke-Elder and Leigh (1965), reviewing the literature, stated that this complication was a rare development of scleroderma, and Stucchi and Geiser (1967) did not report it in their series. However, in the present series no less than seven of the 23 patients with PSS developed this complication, indicating that its incidence may be higher than was previously supposed. In contrast, Duke-Elder (1952) also reported that lid involvement might cause difficulty in lid closure resulting in ectropion with a secondary keratitis. Although fifteen of the present cases had tightening of the lids, these further complications were not observed.

KERATOCONJUNCTIVITIS SICCA WITH XEROSTOMIA AND ARTHRITIS

Sjögren's syndrome is a well-documented manifestation of systemic sclerosis (Shearn, 1960; Tuffanelli and Winkelmann, 1961; Rodnan, 1963). The present series confirms this finding, two out of the 23 patients having a well-defined Sjögren's syndrome. This incidence (9 per cent.) seems to be rather higher than in other series.

CATARACT

The presence of cataract in association with PSS is one of the more difficult findings to evaluate objectively. Some workers have indicated that cataract is a very rare finding indeed in connexion with PSS (Leinwand and others, 1954; Stucchi and Geiser, 1967), but Hartmann and others, (1948), Thygeson (1963), and Sanders (1966) have stated that cataract is among the main ocular manifestations of scleroderma. The present findings incline towards the first opinion, for only one of this series had a mature cataract—and this was probably of the senile type.

FUNDUS

Significant fundus lesions were not observed in this survey though cytoid bodies have frequently been reported (Agatston, 1953; Nelken and Michaelson, 1961; Pollack and

Becker, 1962; Goder, 1964; Manschot, 1965). Their significance is difficult to evaluate since they have been seen in the terminal stages of PSS and in patients with hypertension and renal disease.

FUNCTIONAL IMPAIRMENT

Impairment of vision was not a notable feature in the present series. The eye manifestations appeared to be limited to the more superficial tissues, giving rise to lid involvement in 83 per cent, reduced tear secretion in 48 per cent., keratoconjunctivitis sicca in 30 per cent., and shallow fornices in 22 per cent.

Summary

The ophthalmic manifestations of progressive systemic sclerosis are reviewed in a controlled study of 23 patients. Tightness of the lids, diminished tear secretion, keratoconjunctivitis sicca, and shallow fornices were specific ophthalmic manifestations of the condition. Other forms of eye disease were occasionally seen, but lenticular and retinal changes and functional impairment of vision were not features of this systemic disease.

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