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Ocular involvement in scleroderma

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SUMMARY Thirty-eight patients with scleroderma (progressive systemic sclerosis) without renal failure were subjected to detailed clinical ophthalmic assessment. Abnormalities were frequent. However, many of these, including lens opacities, vitreous frosting, and arteriosclerotic changes, were considered to be age-related, and there were various incidental changes. Posterior subcapsular lens opacities in one patient were probably corticosteroid-induced. Changes related to scleroderma included eyelid abnormalities (stiffness or tightness in 11, telangiectasia in 8), deficient tear secretion (14 cases), and conjunctival abnormalities (injection 19 cases, vascular sludging 27 cases). Iris light reflux (6 cases) was possibly related to scleroderma. The changes related to scleroderma occurred in the 3 types classified according to the extent of skin involvement.

Scleroderma with systemic involvement (progressive systemic sclerosis) is an unusual but not rare disease. Its clinical and laboratory features have been analysed in some detail in numerous papers, reviewed in a recent monograph.[1] Ocular involvement in scleroderma is widely recognised,[2-4] but the nature, frequency, and relevance of the ocular changes remain the subject of some uncertainty. The occurrence of severe fundal changes in acute terminal renal failure associated with scleroderma has been well documented.[5] The purpose of this paper is to explore the clinically significant ocular features in a group of patients affected by scleroderma of different degrees of severity but not in renal failure.

Patients and methods

The study was conducted on 38 outpatients with scleroderma, comprising 31 women and 7 men, ranging in age from 22 to 76 years, median age 60 years. With the exception of one Arabic Lebanese woman all were of Caucasian stock. They were classified according to the extent of the skin sclerosis as proposed by Barnett and Coventry[6] as follows: type 1, skin changes of fingers only (17 patients); type 2, skin changes beyond fingers but more marked in limbs and face (16 patients); type 3, diffuse changes involving trunk (5 patients). Types 1 and 2 are sometimes combined as 'acrosclerosis'. Almost all had evidence of some systemic disorder either clinically or on tests.

The patients were subjected to a careful ophthalmic assessment, including applanation tonometry, biomicroscopy of the anterior segment and vitreous, and examination of the ocular fundus by direct and indirect ophthalmoscopy after the instillation of mydriatic drops.

We sought sludging or segmentation of the blood columns in conjunctival vessels mainly of venular calibre, using biomicroscopic magnification. The appearance of the vitreous fibrils was assessed minutely by biomicroscopy and contact lens examination, with particular reference to thickening, nodularity, or cellular deposits in the vitreous framework. Stiffness and loss of elasticity of eyelid skin was assessed subjectively. Lacrimal function was assessed by examination of the cornea and conjunctiva after the instillation of Rose-Bengal drops and by a Schirmer test,[7] wetting of the filter paper strip less than 10 mm after 5 minutes being taken as indicating reduced tear secretion.

Results

Eyelid changes. Tightness of the skin of the eyelids was found mainly in type 2 cases. Associated shortening of the interpalpebral fissure as reported by Kirkham[3] was found in only 1 patient. Telangiectases of the eyelid skin were generally tiny, seldom exceeding 1 mm in diameter, and were best detected under biomicroscopic magnification.

Lacrimal function. Deficient tear secretion was

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found in a proportion of patients of all 3 types. As some patients with disturbed lacrimal function show normal results at one visit and abnormal results at another, suggesting intrinsic variation or perhaps some influence of environmental conditions, the incidence of tear defects is probably underestimated in this study.

Conjunctival changes. These comprised vascular congestion, telangiectasia and varicosities of conjunctival vessels, intravascular sludging, and loss of fine vessels, all of which occurred in all 3 types. The varicose vessels were found mainly in relation to the corneoscleral limbus. Both varicosities and sludging were greatly reduced by the instillation of a vasoconstrictor solution (phenylephrine HCl 10%) used to facilitate mydriasis.

Lens opacities. Lens opacities mainly of the nuclear or cuneiform type occurred in several patients.

Vitreous changes. Vitreous syneresis and collapse were present in the expected proportion for this age group. Particular attention was paid to the appearance of fine nodularity or encrusting of vitreous fibrils ('vitreous frosting') as a possible index of low-grade intraocular cellular inflammation. It was observed in about one-third of the patients.

Fundus abnormalities. Retinal arteriosclerosis (indicated by variations in calibre, tortuosity or shortening, wall changes in retinal arteries, and arteriovenous crossing changes) was found in several patients of each type. Colloid bodies (drusen) distributed irregularly about the posterior pole were present in all disease types but were not present in any patients below the age of 63 years. The single instance of peripheral schisis of the retina was seen in 1 eye of a 63-year-old patient with retinal arteriosclerosis, and is to be regarded as of senile rather than of dystrophic significance. Cystoid retinal change observed in 1 eye of 1 patient was of uncertain pathogenesis, but might represent a previous localised vascular or inflammatory disorder. (No history of trauma was elicited.) A retinal branch vein occlusion seen in 1 patient was associated with retinal arterial sclerosis with constriction of the affected venous branch due to sclerosis of the adjacent arterial adventitial sheath.

Iris reflux. Spotty areas of transillumination in all zones of the iris caused by localised defects in the iris pigment epithelium were seen in several patients, mainly with lightly pigmented grey or blue irides.

Intraocular pressure. In both patients with raised intraocular pressure the condition was not associated with optic disc changes or visual field loss, and occurred in the presence of a normally open anterior chamber angle.

Discussion

The retinal changes in scleroderma with advanced renal failure have been described[5] but are not applicable to the patients in this study. Other writers have described the following changes in scleroderma without renal failure: eyelid sclerosis and telangiectasis,[2 4] conjunctival congestion or oedema,[2] defects of lacrimal secretion,[2 3] iris atrophy, and retinal exudates.[4] However, no systematic study of ocular changes in a large number of patients as presented has been reported previously.

In considering the large variety of ocular abnormalities revealed in this study some separation of the changes may be made provisionally into those related to the age of the individual, those due to previous or current treatment, and those due to scleroderma or associated disease.

Presumed age-related abnormalities. Certain of the abnormalities—cuneiform and nuclear lens opacities, vitreous frosting (the 'senile peppering' of Koby[8]), colloid bodies of the posterior pole, and retinal arteriosclerosis, are age-related, and their incidence would not appear to differ markedly from that in the general population. Raised intraocular pressure, found in 2 of the 38 patients, is consistent with a spontaneous partial manifestation of chronic simple glaucoma in a middle-aged and elderly population. One of these patients had been on systemic steroid therapy, but steroid provocative testing failed to support the possibility of a steroid-induced glaucoma.

Changes presumably related to treatment. Posterior subcapsular lens opacities were noted in 1 patient who had been treated with systemic corticosteroids for many years. Her lenses were later extracted with a good visual result.

Changes probably related to scleroderma. The following changes, not explicable by age or treatment, were probably related to scleroderma: eyelid changes, conjunctival changes, lacrimal disturbance. The iris changes in the absence of other known causative factors (diabetes, moderate or high myopia, pseudoexfoliation of the lens capsule, and pigmentary glaucoma) are possibly related to scleroderma. The eyelid features (tightness, telangiectasia, and blepharitis) were more common in types 2 and 3, in which the skin changes were more extensive.

Relationship of changes to other manifestations of scleroderma. Two of the most prominent features of scleroderma are alterations in connective tissue and vascular changes. The stiffness of the eyelid skin is in accordance with other connective tissue involvement. The telangiectasia of the eyelids and conjunctiva is probably related to similar changes

seen elsewhere, particularly in the face and hands. The dilatation of venules and intravascular sludging in the conjunctivae are similar to changes described in the nailbed capillaries.[9] It is of interest that the vascular changes are not necessarily greatest in the location of the most marked connective tissue changes, which indicates that these 2 prominent features do not necessarily run parallel in severity. The pathological basis of the decreased lacrimal secretion is not clear, but in the absence of other features of Sjögren's syndrome, except in 1 patient would seem to be specifically related to scleroderma. As is the case with other systemic disturbances in scleroderma,[10] the associated eye changes occur in all 3 types.

Conclusion

Ocular changes which appear to be specifically related to scleroderma include: telangiectasia and dermal sclerosis of the eyelids; a tear defect of varying severity; injection, telangiectasia, and sludging of the blood column in conjunctival vessels; possibly punctate defects of the iris pigment epithelium. Posterior subcapsular lens opacities in 1 case were attributed to long-term systemic steroid therapy.

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