CASE NOTES

SCLERODERMA OF THE CORNEA*

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SCLERODERMA is a rare disease which produces thickening of the skin and subcutaneous tissue followed by atrophy. There is endo- and perivascularitis with an increase in fibrous tissue, and the elastic tissue may be increased or there may be atrophy following hypertrophy. It is usually divided into two types: generalized and circumscribed. The circumscribed type sometimes retrogresses spontaneously.

There have been cases in which the lens (ectodermal in origin) has been attacked, but there is no recorded case of scleroderma of the cornea, the epithelium of which is ectodermal, and for this reason the following case is of considerable interest.

Case Report

An aboriginal bush worker, aged 22, born in New South Wales, and residing at Brewarrina, had two sojourns in Waterfall Sanatorium for treatment of pulmonary tuberculosis. During his second stay, he was suffering from pulmonary tuberculosis, circumscribed scleroderma of the left frontal region, and keratitis in the left eye.

Clinical History

Pulmonary and Skin Lesions.—He was first admitted to Waterfall Sanatorium on March 24, 1948, at the age of 15 years. His mother and his mother's sister had died from pulmonary tuberculosis, and his sister had had pulmonary tuberculosis. As a contact he was x rayed at Brewarrina Hospital, and radiological evidence of tuberculosis was found.

No tubercle bacilli were ever found in his sputum. He was treated by rest in bed only, and on November 28, 1951, was considered well enough for discharge.

He returned to work and had periodical x rays. His health was satisfactory until May, 1954, when he noticed fatigue, shortness of breath, loss of weight, and night sweats. He was admitted to the Dubbo Hospital and treated with streptomycin and P.A.S. from May to October, 1954. He was then admitted to Waterfall Samatorium on October 6, 1954; the sputum was negative and the erythrocyte sedimentation rate (Westergren) 38 mm./hr.

The Mantoux test (0.T.1/1,000) on November 1, 1954, was positive. Repeated tests of the sputum for the presence of tubercle bacilli with culture and guinea-pig inoculations and similar tests of the bronchial secretions, after bronchoscopic aspiration, were negative. There is no record, therefore, of tubercle bacilli ever having been obtained from this patient.

Radiologist's Report (October 24, 1954): "Linear-nodular markings throughout the whole of the left lung field and widespread but less extensive markings throughout the

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right lung field. No definite cavities seen but there is a suspicion of cavitation in the left upper and left lower zone".

The patient was referred to a dermatologist at the end of October because of a skin lesion involving the left half of the forehead. The diagnosis was: "Early hemifacial scleroderma. There is no evidence of tubercular involvement". Therapy with vitamin B complex, vitamin A, etc. was recommended, and the diagnosis of scleroderma was confirmed by biopsy.

A diagnostic thoracotomy was performed for the purpose of lung biopsy on February 21, 1955. Part of the pleural surfaces were adherent at the side and some small areas were removed from pleura and adjacent lung. The report on section was: "Caseation with surrounding epithelioid and fibroblastic reaction. Typical giant cells and focal tubercles are absent but the histological features are almost certainly those of tuberculosis. It has not been possible to demonstrate acid-fast bacilli in the paraffin sections".

Tonsillar biopsy on December 13, 1954, showed: "A simple inflammatory hyperplasia with hypertrophy and ulceration of the surface epithelium. No evidence of more sinister disorders".

X rays of the bones of hands and feet showed no evidence of sarcoidosis.

Progress.—The patient showed clinical and radiological improvement of the chest condition since admission, the treatment being rest in bed only. He had no chemotherapy other than that given before admission.

He left the sanatorium at his own request and against medical advice on July 26, 1955.

Corneal Lesions.—In October, 1954, simultaneously with the development of scleroderma of the left forehead, the patient began to complain of a sore left eye. In November, 1954, a group of small, white, circular, woolly opacities were seen to be scattered about the lower half of the cornea, the general appearance being that of a group of little snowflakes, raised on the surface of the cornea. Some of the patches stained with fluorescein. There was a little infiltration in the substantia beneath and surrounding the patches. There was not much ciliary flush.

The patient complained of lacrimation and photophobia, but the degree of distress was slight compared with the number of the corneal lesions. Atropine ointment 1 per cent. three times daily and aureomycin ointment four times daily were ordered with pad and bandage.

Within a fortnight the snowflake nodules first seen had gone, leaving shallow ulcerations with mild surrounding keratitis. But as the old snowflakes ulcerated, showing little tendency either to heal or progress, fresh little deposits appeared, some of them in the upper half of the cornea.

Treatment was continued as above, the lesions continuing with little change; as some lesions slowly healed, the new snowflakes disintegrated and went through the same process as their predecessors, the disease gradually spreading over the upper cornea as it had over the lower. At no time was there much vascularization or irritation, the distress being disproportionate to the apparent corneal disturbance. Moreover, there was no sign of involvement of any deeper structures, iris, aqueous, lens, vitreous, and retina remaining beautifully clear.

At this chronic stage, Dr. Conrad Blakemore kindly examined the patient in consultation, as it seemed possible that the corneal disease was in fact scleroderma. He felt that there was not sufficient evidence to warrant a definite diagnosis, and advised a continuance of the previous treatment as the pathological process was confined to the more superficial half of the cornea and the eye was reasonably quiet. By February, 1955, a definite recession was occurring, the number of staining ulcers was reduced to one, and in the older portions the amount of infiltration was less.

Simultaneously, the scleroderma of the left frontal region was beginning to fade and shrink, so that one could watch the lessening of the lesion in the forehead at the same

time as an obvious improvement occurred in the cornea. The indolent healing proceeded over a period of more than 2 months until mid-April, 1955, when the last ulcer had healed. Cortisone drops 0.5 per cent. four times daily were added to the therapy and the atropine was discontinued. A diminution of corneal infiltration now slowly followed, and the facial scleroderma practically disappeared, the patient's dark skin making it difficult to date its final disappearance.

By July 26, 1955, when the patient left the sanatorium, the scleroderma of the forehead had disappeared except for a slight line of demarcation and a very faint pallor, while the eye was white and quiet with good vision, in spite of some residual cloudiness of the substantia. The patient was illiterate and monosyllabic in conversation, so that it was difficult to assess his subjective condition.

Summary

While at Waterfall Sanatorium, the patient was suffering from pulmonary tuberculosis, and meanwhile, in October, 1954, he developed scleroderma of the left frontal region extending to the root of the nose, with simultaneous lesions of the superficial portion of the left cornea. Both conditions continued in an indolent manner until February, 1955, when they showed signs of healing, and by July, 1955, both had completely healed.

In view of the ectodermal origin of the corneal epithelium, the juxtaposition of the skin and corneal lesions, and the concurrent nature of the two conditions, one is drawn to the conclusion that the corneal lesions were manifestations of scleroderma. Dr. Conrad Blakemore concurs in this opinion.

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