

Two Cases of Scleromyxedema

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Abstract. We report two cases of scleromyxedema treated with melphalan (alkeran) and dermabrasion, both patients with central nervous system involvement. In the first case, herpetic encephalitis, possibly due to the immunoglobulin disturbance, preceded the skin changes. In the second case, impaired cerebral function was caused by a meningeoma. In the first case of short duration it seems as if progression of the disease has been arrested. In the second, of more than 26 years duration the skin changes have seemed unchanged, sclerotic for some years, though some improvement was noticed following dermabrasion.

Key words: Scleromyxedema; Melphalan (Aleran); Dermabrasion; central nervous system disease

Scleromyxedema (lichen myxedematosus or papular mucinosis) is usually a slowly progressing connective tissue disease with characteristic skin changes, consisting of dense groups of lichenoid papules and extensive thickening of the skin in the affected areas. A striking feature is the deeply furrowed glabella. Several reports during the last 10–15 years indicate that many patients have had systemic complications, the most frequent being a paraproteinemia (2, 3, 4, 5, 8) whose relationship to the skin manifestations is unexplained, just as the skin changes themselves are of unknown etiology.

Histopathology shows an accumulation of mucopolysaccharides in the dermis stained blue with alcian blue, and proliferation of large stellate elongated fibroblasts (1, 7). Electromicroscopy shows the dermis to be dominated by collagen fibrils and an accumulation of peculiar connective tissue cells, while elastic tissue is sparse and in some areas completely absent (1). Recently, synthesis of a paraprotein identical with the paraprotein in serum has been demonstrated in the skin lesions (5).

Contradictory autopsy reports describing deposition of acid mucopolysaccharides in internal organs have appeared. McCuiston (6) presented a case with mucinous material in connective tissue and vessels throughout many organs, while Rudner (7) reported two cases without any deposition of mucopolysaccharides.

Cerebral involvement of the disease has been mentioned in a few cases (6, 7). The present study describes 2 cases with disease of the central nervous system.

CASE REPORTS

Case one

A 41-year-old woman was admitted to our Department in July 1977 with an eruption of 6 months' duration appearing on the face, hands and the extensor sides of the forearms. In April 1975 this previously healthy woman had been admitted to a neurologic department because of a serious herpetic encephalitis. In September 1975 a paraprotein located in the IgG region was discovered, both in the serum and in the spinal fluid.

On admission to our Department the diagnosis scleromyxedema was evident. The patient was started on melphalan (alkeran) (2) 5 mg a day for 10 days, followed by 2 mg/day. Because of a leukopenia and thrombocytopenia this dose was further reduced in December 1977 to 1 mg/day. Upon rehospitalization in November 1977 her skin changes on the face had become nodulocystic (Fig. 1), and additional changes could be seen on the shoulders and thighs. The treatment was continued. During the next few months, however, her skin manifestations slowly improved and this has been followed up by dermabrasion of the face (Fig. 2). She still has significant skin changes on the arms, shoulders, buttocks and knees—though less evident than before—interspersed with scattered areas of normal skin. The treatment with alkeran 1 mg/day continues.

Case two

A 65-year-old woman was transferred from the Department of Internal Medicine to our clinic in May 1978. She had a skin disease of about 26 years' duration. In 1973 she received the diagnosis scleromyxedema. For half a year she was treated with penicillamine (dimethylcystein), but stopped the treatment herself.

When we first saw the patient the greater part of her trunk and the extremities were covered by erythematous, infiltrated skin, turning partly sclerotic in the antecubital and popliteal spaces. From the borderlines of these areas multiple keratotic infiltrated streaks stretched, with the appearance of Koebner phenomena. Her face was stiff, with many sclerotic pale red noduli. She could not close her eyes, was troubled by a constant flood of tears and had a microstomia which impeded her eating (Fig. 3). An IgG paraprotein had previously been found and was now confirmed. Moreover we found an impaired cerebral function and further investigations and subsequent surgery revealed a benign meningeoma. She, too, was started on melphalan (alkeran) 5 mg/day for 6 days, followed by 2 mg/day for 2 weeks. The treatment was temporarily discontinued during her stay at the neurosurgical department, but reinstated on her return. It was withdrawn after a further 3 weeks' treatment due to a drop in her leukocyte count and has not been restarted yet. Dermabrasion of the glabella has been carried out (Fig. 4) with excellent results.

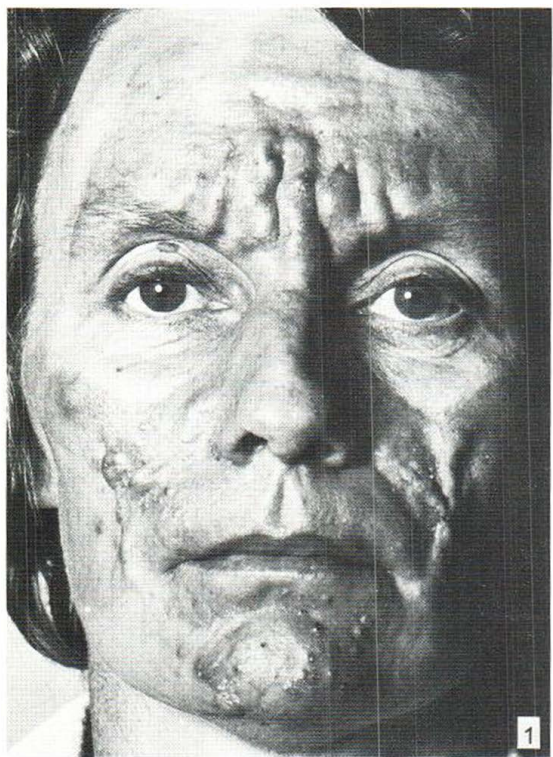


Fig. 1. Patient (case one) when her skin changes were at their most serious.

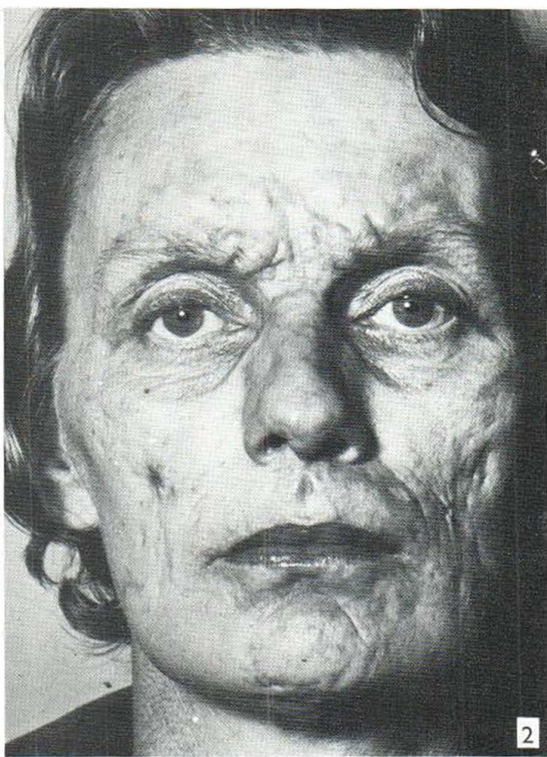


Fig. 2. Patient (case one) after dermabrasion.

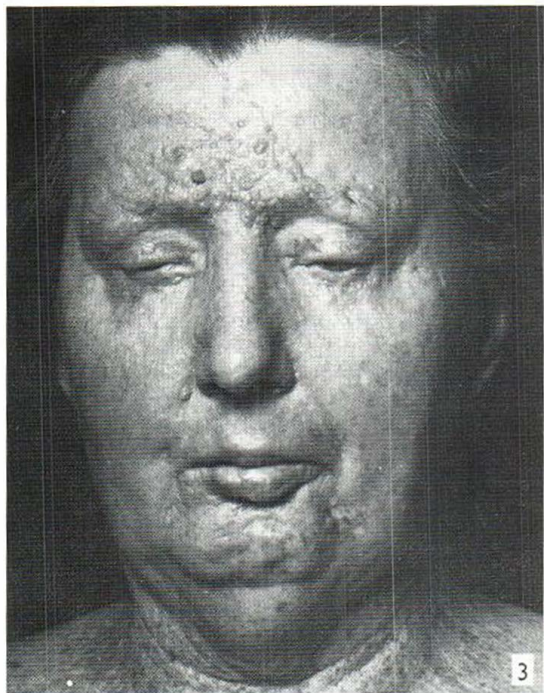


Fig. 3. Patient (case two) before dermabrasion.



Fig. 4. Patient (case two) after dermabrasion.

DISCUSSION

We report two cases of scleromyxedema with cerebral disease, treated with alkeran and dermabrasion. These add support to previous data (6, 7) indicating an increased risk of central nervous system disease in scleromyxedema. The first case presents a firm diagnosis of herpes encephalitis, which makes suspicion of cerebral involvement caused by scleromyxedematous plaques less likely. Immunoglobulin disturbance, with paraprotein formation might, however, have helped to lower the patient's resistance to the herpes infection. In the second case, we must so far assume that we are dealing with a coincidence of two diseases, but it is noteworthy that the histologic appearance of a meningioma is dominated by large stellate fibroblasts, just as found at the skin microscopy.

Available literature concerning effective treatment of scleromyxedema is sparse. Feldmann & Shapiro (2) reported a case in 1969 with a dramatic response to melphalan (alkeran), and Wright & Franco (8) treated a case successfully with alkeran and dermabrasion, while Hill & Crawford (4) used radiation therapy.

In our first patient we were able to start treatment with melphalan (alkeran) and later dermabrasion, before the skin had turned completely sclerotic around the natural openings of the face. At present it seems that progression of the disease has been arrested. The existing sclerotic areas in the glabella and on the cheeks have been and will be further treated with dermabrasion. In the second patient it is hard to say if there has been any effect of the treatment with alkeran. Treatment has only proceeded systematically for a very short time. Dermabrasion has had some beneficial effect.

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Absence of Cataract Ten Years after Treatment with 8-Methoxypsoralen

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Abstract. Thirteen patients treated with 4-41 g of 8-methoxypsoralen (8-MOP) for vitiligo were examined with regard to ocular damage 2-12 years after start of the treatment. No signs of eye damage were registered. This result indicates the need for more comprehensive retrospective examinations of 8-MOP-treated patients in order to gain information on the practical risk of cataract formation.

Key words: Cataract; 8-methoxypsoralen

The risk of cataract formation during photochemotherapy (PUVA) is a source of concern to an increasing number of dermatologists. However, cataracts after PUVA treatment have been observed only in experimental animals treated with PUVA for prolonged periods with very high doses of psoralens (2, 3). The UVA light penetrates both the cornea and the aqueous humor, i.e. it reaches the lens, also in the human eye (1). 8-methoxypsoralen (8-MOP) has been found both in aqueous humor and lens after oral administration (6). The physical prerequisites for a photochemical reaction thus are available also in the lens of the human eye. Despite this fact, long-term use of psoralens for the treatment of vitiligo in many patients does not seem to have caused any cataracts (4). However, no adequate follow-up studies with ophthalmologic examination of treated patients seem to have appeared as yet.

PATIENTS AND METHODS

Mainly during the 1960s, patients with vitiligo were treated orally with 8-MOP, Meladenine®, 10 mg *t.i.d.* in