SCLEROMYXOEDEMA

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Scleromyxoedema (Arndt-Gottron) is a clinical and histological entity which has been brought into prominence lately.[1-3,7] It is not considered to have any connection with thyroid disease or scleroderma, and the name should therefore not call up any such associations. It is closely related to, and possibly identical with, conditions variously called papular mucinosis, lichen myxoedematosus, and lichen fibromucinoidosus.[5] Possibly some eruptive collagenomas properly belong here. An obscure cerebral disorder was noted in our patient, which may have some pathogenetic significance, particularly since marked cerebral and psychological symptoms have been noted before by others.[2-4]

CASE REPORT

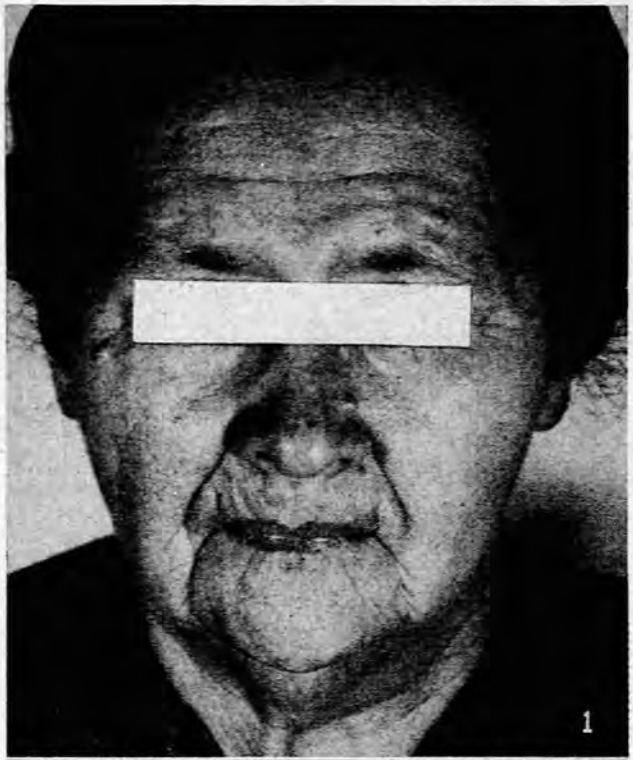
A married housewife of 64 was seen in December 1958 for a disfiguring swelling of the face and extensor surfaces of the forearms which had lasted for the preceding 4 months. It began while she was convalescing from an obscure cerebral disorder which may have been an encephalitis. It started with a cold and severe headache, followed by maniacal confusion and coma. No vascular cause could be established for it at the time. The patient recovered in a few days from the coma and later seemed normal but for a little residual incoordination in the right hand.

The skin changes began with attacks of swelling on the face and forearms, which subsided only partially between the acute phases. The residual skin thickening became darker and scaly on its surface and showed temporary scarlatiniform and urticarial flushing in the acute episodes. The swellings were provoked by heat but not by light, and the skin stiffness was marked in the mornings, when she would try to massage some of it away. There were no other symptoms. At first she also had some similar swellings round the waist and on the ears and gums but these did not last long. The legs were unaffected until the skin changes had been established for about 8 months, and then the anterolateral surfaces of the calves became diffusely involved. Some loss of hair was also noted.

When the patient was first seen the forearms, particularly on the extensor surfaces near the elbow, were brownish and leathery and would only fold in big folds with no smaller wrinkles (Fig. 2). Towards the flexor aspect they were less leathery and the infiltration faded into a fine colourless micropapular fixed eruption (Fig. 3) with a linear or moniliform arrangement in some of the groups of micropapules. The facial skin (Fig. 1) showed a similar parchmenty surface with palpable thickening, and some transient reddish 'angioneurotic' swelling of the upper lip. Elsewhere the skin was normal. There was no evident loss of hair.

General examination showed a hypertension with no tachycardia, a left axis deviation, and no demonstrable renal disorder. There was a pea-sized nodule attached to the thyroid, but thought to be separate from it. It has not been explored as yet. A tracer dose of radio-active iodine showed a hyperthyroidism on one test alone. The basal metabolic was +5, and the protein-bound iodine test gave 6·9 μg %. Blood cholesterol, twice determined, was in the region of 300 mg. %. The total blood proteins were 6 g. % (albumin 38·5%, α_1-globulin 7·7%, α_2-globulin 11·0%, β-globulin 14·3%, γ-globulin 28·5%).

The *histological material* comprised 4 biopsy specimens fixed in formalin, from the diffuse and papular lesions of the forearms. Sections were stained with H and E, toluidin blue, thionin, PAS, Hale's iron technique, mucicarmine, silver for reticulin, elastic, PAH and trichrome. The *epidermis* showed marked pigment increase in the basal layer, some effacement of the papillae, no atrophy, a little intracellular vacuolation, and a thick stratum corneum. The *dermis* (Fig. 4) showed all the important changes in its outer half, viz. a loose oedematous and fibrillar thickening in the isolated lesions and a denser thickening in the more diffusely thickened areas. The papular lesions consisted of a fine-spun collagen meshwork arranged in whorls and criss-cross patterns. The some-



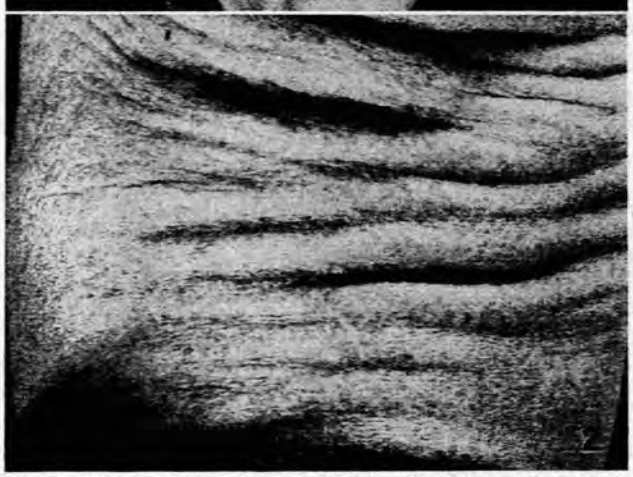


Fig. 1. Scleromyxoedema (female aged 64). Whole face thickened and leathery, with accentuated pleating round mouth.
Fig. 2. Scleromyxoedema. Portion of forearm skin showing thick loose folds in infiltrated area. Skin dark and scaling.

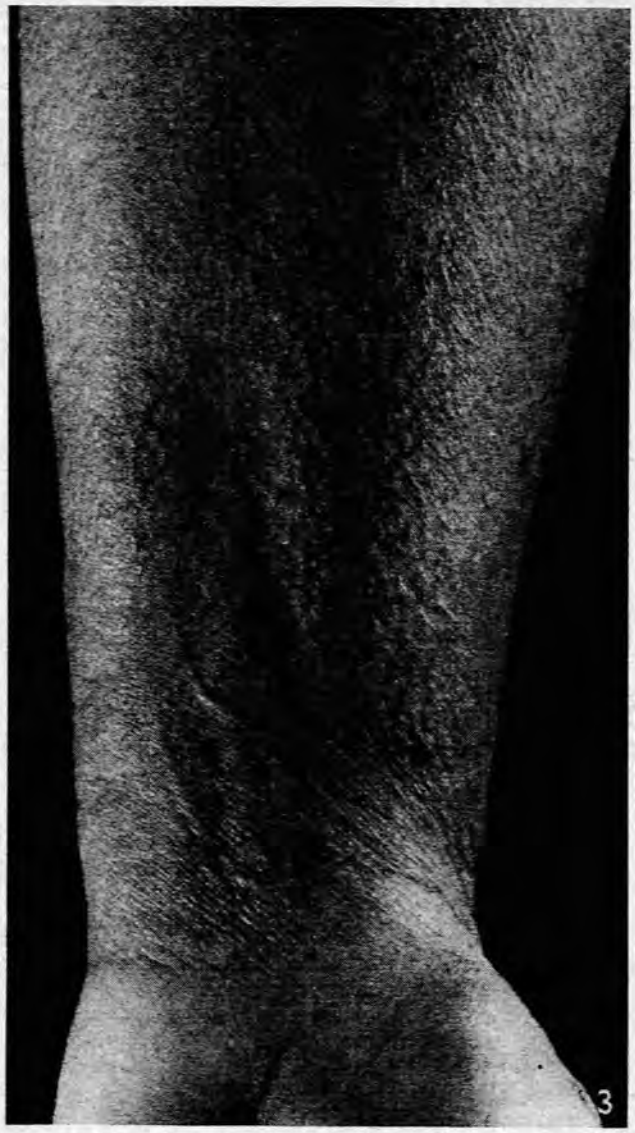


Fig. 3. Scleromyxoedema. Moniliform and reticulated arrangement of colourless micropapules on flexor aspect of forearm.

what larger collagen bundles between this meshwork showed irregular staining, with many pale, shredded, amorphous and foggy bundles. Thick collagen bundles tended to show fewer of these changes and stained in a normal way throughout. In the fine meshwork, fibroblasts were plentiful, and some of these showed a hazy cloud of faintly basophil material around them merging with the collagen. Pericapillary spaces were enlarged and showed an increase of adventitial, fibroblast and lymphocyte cell types. These pericapillary cells often showed vacuoles which indented the nuclei but no stainable material was contained in them. Mast cells were present, but in the minority compared with the fibroblasts. Elastic was present, but degenerated and somewhat reduced in amount. The pericapillary and intercollagenous interstitial spaces (Fig. 5) were partly empty, being created to some extent by tissue shrinkage but probably also by interstitial fluids which were lost in fixation and processing. These spaces were filled by material of a mixed and uncertain type. Granular, fibrillar, filamentous and cloudy membranous structures were seen, which merged variably with the surrounding fibroblast cells and dermis fibres. Some fibrils, but not all, stained like reticulin. Some of the material could have consisted of small collagen fibres staining atypically. Elastic was absent. There was streaky and granular Schiff positivity with PAS staining in the interstitial areas. Very mild metachromasia was noted, and mucicarmine stains showed nothing definite. Hale's dialysed iron technique showed blue fibril staining resembling reticulin fibres.

DISCUSSION

Keining and Braun-Falco[3] believe that scleromyxoedema represents a strong dermal fibroblastic reaction which progresses only in an imperfect degree to fibrosis, and that the stimulus to it is the abnormal transudation of altered serum proteins into the tissue spaces, where they form a complicated conjugation with a variety of tissue mucins. Their arguments are based mainly on the histochemical behaviour of the mucins

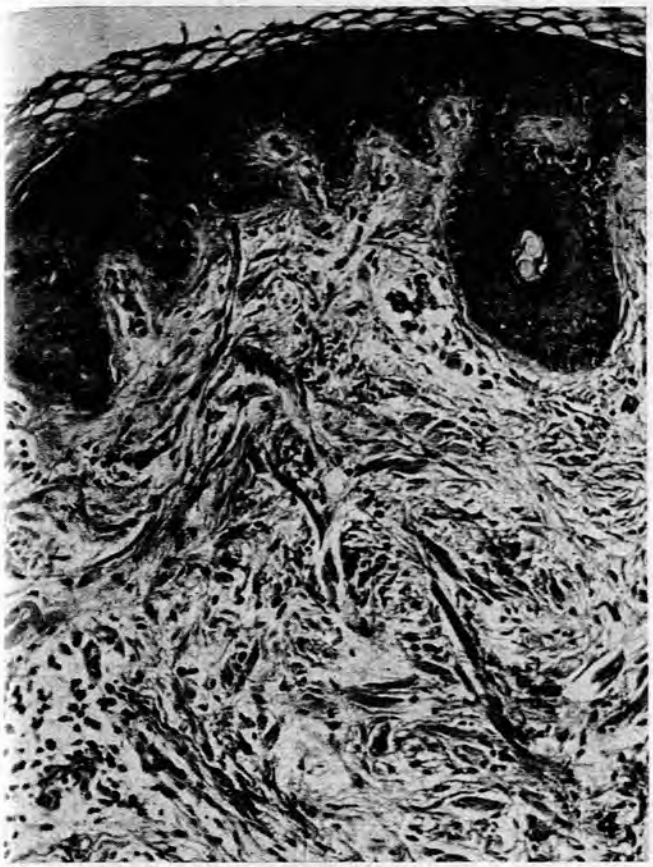


Fig. 4. Scleromyxoedema. Low power of the dermis (see text).

Fig. 5. Detail of the fibrillar and interfibrillar alterations (see text).

in the general clinical setting, and accord well with our histological evidence.

However, the lack of abundant stainable mucin in our case presents an obstacle. Three factors may contribute, viz. formalin fixation, random and uncontrollable variations in mucin staining reactions, and a genuine lack of mucins. In this connection, 2 recent publications report cases which are typical of scleromyxoedema in all respects apparently, but their authors attach entirely different interpretations to the changes, and the resemblances to scleromyxoedema have evidently escaped them. Thiers *et al.*[6] stress the cellular proliferation of fibroblasts with inhibited formation of reticulin and collagen, an increase of elastoid (orcein-staining) material and increase also of an interstitial granular basophilic material. Nicolau and Balus[8] emphasize the reticular to fibroblast increase and the connective-tissue degeneration. The former authors call the disease a fibro-elastoidosis and the latter a benign reticular-fibroblastic cutaneous reticulosis. Difficulties with mucin demonstration have been indicated recently[5,9] in this disorder. They recall the description by Dubreuilh in 1906 of this disease as miliary fibromas with subsequent scleroderma (quoted[2]).

The remaining feature worth discussing is the clear relation in our case between a cerebral attack and the onset of symptoms. Although our patient was psychologically normal, apart from a poor memory, in the only published autopsy in scleromyxoedema[4] focal demyelination and gliosis were reported in the brain. Nevertheless, we have no evidence of how the brain disorder, whatever its nature may be, could be related to scleromyxoedema. There is no evidence that it is due to a primary cerebral vascular disease with mucin deposits in the vessels, nor is there any indication as yet of a hypophyseal-hypothalamic mechanism.

SUMMARY

A typical case of scleromyxoedema (Arndt-Gottron) is described. Theories on the pathogenesis have been noted, even where the disease has been misdiagnosed, and the significance of organic cerebral disorder in this disease is indicated. In the case presented, the fibroblastic and connective-tissue alterations were more prominent than alterations in the mucin content of the skin.

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