DERMATOMYOSITIS IN THE TRANSVAAL AND ITS OCCURRENCE IN THE BANTU*

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It has long been felt that dermatomyositis is an unduly common disorder among the Bantu of the Transvaal. Observant practitioners in the country districts are aware of its occurrence without always being able to give a name to the condition. At the Jane Furse Hospital, Sekhukhuniland, Transvaal, the disorder is well known; 10 cases were seen between February 1958 and June 1963, 4 of which were reported in detail by Horsfall.[1] Since that time a further 5 cases have been seen there, bringing the total to 15 Bantu cases in 10 years. As this hospital serves a population of 200,000 Bantu, the minimum annual incidence of dermatomyositis in this racial group lies in the region of 7·5 cases per million.

Over a 7-year period, 35 Bantu and 4 White cases of dermatomyositis were seen by us at the H. F. Verwoerd Hospital, Pretoria—an average of 5 Bantu cases per year. By comparison, in the same hospital, the incidence of acute lupus erythematosus in the Bantu has been found to be 2·4 cases per year.[2] The effective size of the Bantu community served by the hospital has never been estimated, but it probably lies between 600,000 and 1 million inhabitants. It can be seen that this figure for Pretoria gives a frequency of dermatomyositis comparable with that found in Sekhukhuniland. Supposing the hospital at Pretoria served the entire Northern Transvaal Bantu (2¼ million), the annual incidence of dermatomyositis would be not less than 2·1 cases/million.

This minimum figure contrasts sharply with a figure for polymyositis (of which some 50% are examples of dermatomyositis) of 1·2 cases/million/year in Southern California and Northern England.[3] In these two White communities, the figure for polymyositis is therefore about one-fifth of our frequency estimate in the Bantu for dermatomyositis *alone*, making the discrepancy greater still. Even if we ignore the patients who failed to come to hospital and misdiagnoses in those who did, the Bantu incidence figures for dermatomyositis are still 10 times higher than in the White.

Comparing 3 serious 'collagenoses', systemic scleroderma, systemic lupus erythematosus and dermatomyositis, one's impressions of relative frequency in Pretoria among Bantu and Whites are as set out in Table I. Rough annual ad-

TABLE I. RELATIVE FREQUENCY OF LE, SCLERODERMA AND DERMATOMYOSITIS

Race	*Commonest*	*Intermediate*	*Rare*	*Frequency ratio*
White	Acute LE	Scleroderma	Dermato-myositis	(7 : 2·5 : 1?)
Bantu	Dermato-myositis	Acute LE	Scleroderma	(5·5 : 2·4 : 0?)

*Date received: 22 August 1968.

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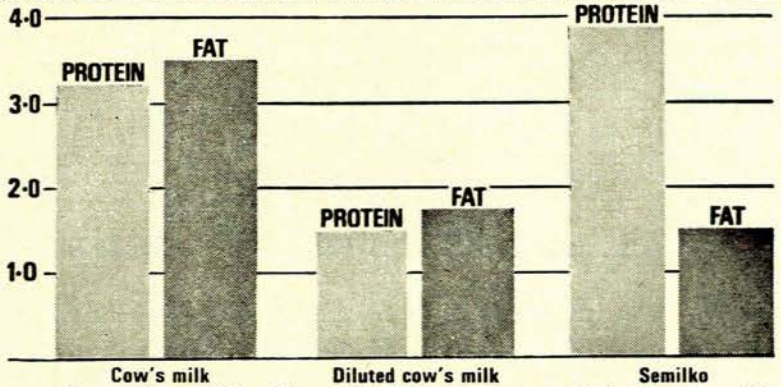

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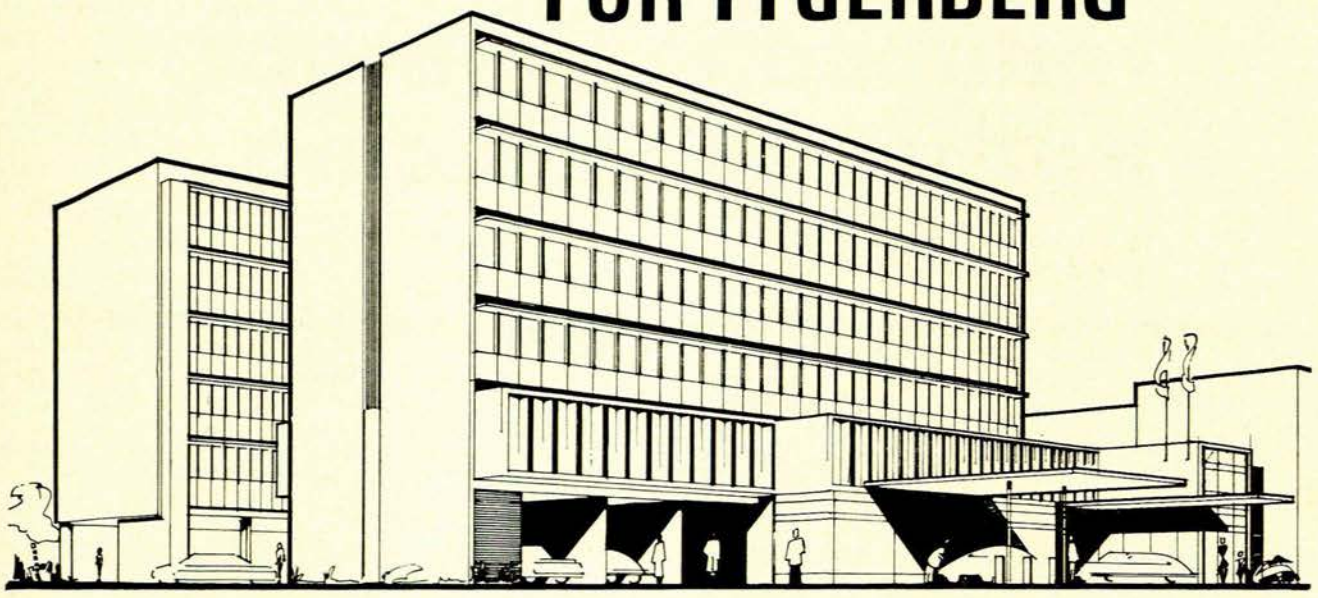

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mission ratios for the 3 disorders in the 2 patient groups are given in brackets.

Comparative figures for Whites are available from Mainz, Germany, where acute LE occurs almost twice as commonly as dermatomyositis.[4] In California, Pearson[2] reports about equal figures for polymyositis and scleroderma, with systemic lupus erythematosus also twice as common as either. In White patients from the Transvaal the figures for scleroderma are increased somewhat by the systemic variety occurring in goldminers with many years of service underground. The peculiarly high frequency of dermatomyositis in the Bantu has evidently not been duplicated elsewhere.

In White populations dermatomyositis may be brought on by carcinoma in the older age-group, and one might expect higher frequency in the Bantu to have some such precipitating cause. Though cases will doubtless appear in which carcinoma brings on dermatomyositis in the Bantu, it is still true, as stated by Marshall,[5] that the association of dermatomyositis and malignancy has not been reported in the Bantu. Furthermore, the Bantu cases of dermatomyositis which boost our series fall in the young adult group before the 'cancer age'. Clearly there must be influences other than carcinoma which alter the frequency, age distribution and precipitating factors of dermatomyositis in the Bantu.

CLINICAL FEATURES

Table II summarizes certain facts on 15 recent cases. Symptoms, which had generally lasted ½ - 1 year, consisted

TABLE II. ANALYSIS OF CASES

Age	*Sex*	*Outcome*	*Assoc. findings*
		Bantu cases	
10	F	Died, PM	—
11	F	Steroid control (full)	—
16	F	Died, PM	Tuberculosis
20	F	Died, PM	—
24	F	Steroid control (full)	Postpartum
24	F	Steroid control (partial)	—
25	F	Steroid control (partial)	Postpartum
31	F	Steroid control (partial)	Unexplained amenorrhoea
46	M	Steroid control (partial, relapsing)	—
56	M	Died, PM	Tuberculosis
70	F	Steroid control (partial)	—
		White cases	
7	M	Died, PM	—
17	M	Recovered	—
51	F	Deteriorating	Breast Ca.
65	F	Died ? anaphylaxis	—

(in order of frequency) of skin rash, asthenia, decreased performance of normal movements grading down to marked weakness and inability to walk, swelling of face and/or neck, joint pains, dysphagia, cold extremities and stomatitis. Clinical findings, in order of frequency, were muscular weakness of the limb girdles, abnormal consistency of the muscles on palpation, miscellaneous skin lesions (ulcers, scaling, pigmentation), difficulty in rising or climbing, oedema of the face and neck, poikiloderma, flexion contractures, asthenic prostration, heliotrope erythema of the face and other erythemas, palatal paresis, alopecia or hypertrichosis, peripheral circulatory insufficiency, muscle tenderness, skin sclerosis and mucosal lesions.

Abnormal laboratory findings from all cases were, in order of frequency, abnormal serum proteins (increased gammaglobulin), suggestive changes in muscle biopsy, a moderately raised ESR, raised serum enzyme levels (aldolase, SGOT, SGPT) and a suggestive skin biopsy. Hospital care on each admission was needed for 2 months on the average, and longer times and readmissions were recorded for those who responded partially or not at all to treatment. Six of the patients have already died, and post-mortem examinations were done on 5.

HISTOLOGICAL FEATURES

Muscle Changes

The following types of change, based on 18 specimens, were fairly characteristic:

Fibrillar change. The myofibrils became separated by spaces and stood out more clearly. The fibres themselves and their nuclei were otherwise little altered.

Granular necrosis. A progressive loss of sarcoplasm and cross-striation was noted, with ultimate breakdown into a fine basophilic material. The subsarcolemmal nuclei sometimes took up a central position in the fibre at this stage.

Eosinophilic necrosis. These fibres were usually enlarged, homogeneous and eosinophilic, though the same changes were occasionally noted in fibres which were normal in size and atrophic. In most of these enlarged fibres, cross-striations were absent and their staining characteristics reminded one of haemoglobin casts in the renal tubules, possibly from excess myoglobin. The nuclei were usually peripheral, much shrunken and pyknotic. Such fibres were inclined to disintegrate through the appearance of fissures and vacuoles.

Phagocytosis by histiocytes was not usually marked, but occurred after both granular and eosinophilic necrosis. After removal of the debris, the muscle cells consisted merely of collapsed and empty sheaths.

Relationships between necrotic changes. It was difficult to arrange the types of necrosis seen into a single chronological sequence. The various changes often occurred close together or even in the same muscle fibre. Thus, fibrillar change did not necessarily precede eosinophilic or granular change, but could proceed side by side with them. Some enlarged and eosinophilic fibres could break up, leaving a granular basophilic residuum. In fibres with granular change, small irregular chunks of eosinophilic material were also seen.

Regeneration. Marked proliferation of the subsarcolemmal nuclei occurred in many fibres. These nuclei were often enlarged, with prominent nucleoli and formed streams or small knots under the sarcolemma. Extensive subsarcolemmal nuclear proliferation was also seen in some relatively normal-looking fibres. Some basophilic regenerating fibres were seen with proliferating subsarcolemmal nuclei, but these had to be searched for.

Inflammatory cell infiltrates. Focal infiltration of chronic inflammatory cells, usually lymphocytes, but sometimes predominantly plasma cells, was seen in the interstitium, often in areas which had undergone fibrosis, but not exclusively so. They were sometimes associated with

necrotic fibres undergoing phagocytosis, but otherwise they were not specifically associated with necrosis of fibres.

Interstitial fibrosis. In muscles where there had been extensive fibre loss, many proliferating and collagen-forming fibroblasts were seen between the surviving fibres. In late cases extensive interstitial fibrosis was seen.

No constant or specific histological change was seen in blood-vessels and neurovascular bundles beyond an occasional moderate oedema of the neurovascular bundles. Hypertrophy of individual muscle fibres occurred here and there, but the large fibres were usually those that had undergone eosinophilic necrosis. The muscles were usually patchily affected and this could be clearly seen in cross-section where only occasional fibres were picked out by the necrotic process. The lesions were also focal within individual fibres and this could be seen in cross-section, a fibre appearing relatively normal undergoing sudden transition to eosinophilic necrosis.

Skin Changes

Thirty-two biopsy specimens were examined. Changes in the epidermal cells comprised mainly atrophy, cell shrinkage, degeneration of nuclei, oedema with vacuolation and granular change of the cytoplasm. Sometimes the tonofibrillar structure stood out more clearly than normal. Pigment incontinence, colloid body formation and fraying-out of the dermo-epidermal junction were noted in the papillary layer. Alterations in the collagen and in blood-vessels were somewhat vague and inconstant. The dermal collagen showed some swelling, granularity, fraying, oedema and at times sclerosis, while the vascular endothelia showed granularity and folding with some swelling of the cells and the surrounding layers, with telangiectasia in places. Cellular infiltrates of lymphocytes and histiocytes were not conspicuous. In some specimens the skin showed evidence of epidermal regeneration. No biopsy material was collected from sites showing clinical signs of ischaemia.

POSTMORTEM FINDINGS

The major postmortem findings are summarized in Table III. In 4 of the 5 cases myocardial changes were present. These changes varied from individual fibre necrosis with loss of striations and nuclei to minor changes involving fibre shrinkage and hydropic changes, resulting in a moth-eaten appearance in the affected muscle.

Inflammatory cell infiltrates were absent in 3 of the cases, but mild lymphocytic and plasma cell infiltrations were present in one.

In one case early interstitial fibrosis was seen.

In one of the cases marked systemic venous congestion was present, which was thought to be due to myocardial failure as a result of the heart muscle lesion.

DISCUSSION

Age Distribution

Even though dermatomyositis can arise at any age, 2 clinical forms which lie on either side of the active reproductive period are recognized in White patients. These forms are the childhood or juvenile type and the 'postmenopausal' type (age 40 - 60 years), of which the former is rather apt to develop calcinosis and the latter is not infrequently associated with carcinoma. In the Bantu, this separation in the peaks of age distribution is not a feature. The juvenile type is readily recognized, and the calcinotic form has also been seen by us, but most cases appear during early and middle adult life, and in the older adult cases no associated carcinoma has been encountered.

Epidemiology

Our first observations on dermatomyositis in the Bantu were made 19 years ago, and were published in 1951.[4] Since then we have noted a periodic tendency to 'clustering' of cases of dermatomyositis in hospital. On several occasions we have had, over the space of a year or two, a singularly large number alternating with practically none over similar periods in between. We have so far no proof that this clustering is more than could be expected from chance alone. It is the only hint, and a questionable one at that, which might relate dermatomyositis to some sort of infectious particle.

Associated Conditions

Does anything suggest a reason for this apparent difference in age distribution and frequency between Bantu and White? Since dermatomyositis is not clearly hereditary, an inherited racial predisposition is unlikely and would require more evidence than we can at present offer to support it. The only other notable and possibly relevant feature of the Bantu population is the high incidence of tuberculosis—14 times more frequent than

TABLE III. POSTMORTEM FINDINGS

Race	*Age*	*Sex*	*Underlying disease*	*Myocardial involvement*	*Heart weight (in G)*	*Other findings*
Bantu	10	F	Nil	Absent	110	Suppurative bronchopneumonia
Bantu	16	F	Tuberculous bronchopneumonia and peritonitis	Mild—present	150	—
Bantu	56	M	Tuberculous bronchopneumonia and enteritis	Present—mild	450	Cor pulmonale with systemic venous congestion
White	7	M	Nil	Present—moderately severe	100	—
Bantu	20	F	Nil	Moderately severe with systemic venous congestion suggesting heart failure	230	Occasional bilharzial granuloma in liver. Terminal bronchopneumonia

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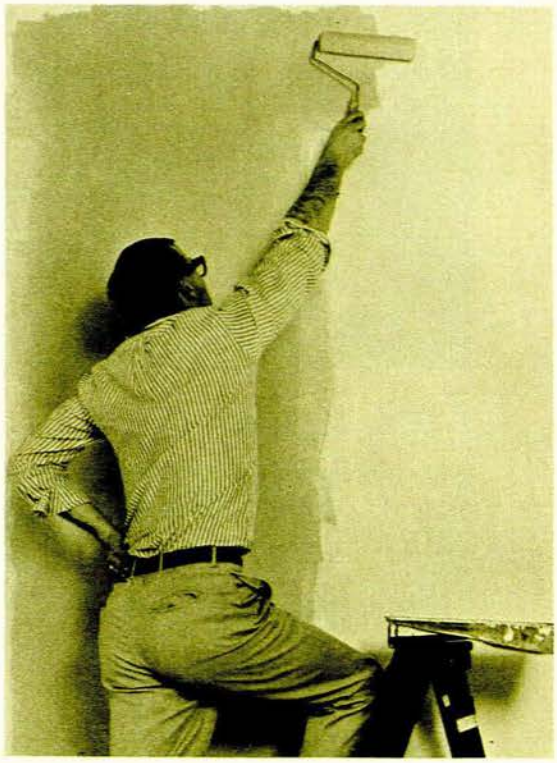

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in Whites. Malnourishment, pregnancy, the puerperium and tuberculous infection may singly or jointly alter the patient's responses and immune reactions. Though it is hardly possible to bring these differences of reactivity into direct relationship with dermatomyositis, a surprising number of case reports refer to latent or overt tuberculous infection in dermatomyositis. Roder and Langer[7] have discussed this feature, and indicated how tuberculosis facilitates antibody formation. Freund's adjuvant—an emulsion of an antigen with extracts of *Mycobacterium tuberculosis* —greatly enhances experimental antibody production by such antigens. Auto-antibody production in dermatomyositis could thus be favoured in a tuberculous subject. Unhappily, this argument fails to prove that dermatomyositis is an auto-immune disease, nor does it say why other auto-immune diseases are not likewise enhanced. It touches nevertheless on an obscure connection which must exist between dermatomyositis and the muscular wasting of carcinoma, malnutrition and tuberculous cachexia.

Relation between Skin and Muscle Changes

We cannot be sure how or why skin and muscle are so often affected in continuity. In the face, where the muscles insert directly into the dermis, erythrocyanotic oedema, poikiloderma and sclerosis are seen in this order of frequency. On the trunk and proximal parts of the extremities the skin is connected to muscle mainly through the perforating neurovascular bundles which traverse the deep fascia. Here the poikilodermas are commonest. Where the skin overlies tendons in the distal parts of the extremities, sclerotic and vascular changes predominate. Whatever the reason may prove to be, it seems at present as if the surface changes are in some way governed by the disease of the underlying muscle, and that the surface expression will depend on the kinds of anatomical connections which exist between the muscle and the overlying skin.

Diagnosis

Failure to diagnose dermatomyositis is often the result of:

(*i*) Failure to recognize the rash, by calling the dermatosis allergic, pellagrous, renal oedema, etc.

(*ii*) Failure to appreciate that the patient is sick from weakness and not weak from sickness. This applies whether the patient is utterly prostrate or merely tardy and hesitant in his movements. Slightly nasal speech is easily overlooked.

(*iii*) Failure to palpate the muscles of the girdles and the extremities and note their altered and uneven consistency.

(*iv*) Failure to recognize the ulcerative or sclerotic skin changes as part of dermatomyositis.

We have ourselves fallen victims to all these sources of error at different times.

When the diagnosis of dermatomyositis seems probable on clinical grounds, the laboratory findings are a useful adjunct, but they have not in our experience ever by themselves pointed the way to a correct diagnosis, nor would we expect them to do so.

Pathogenesis

Changes seen with the light microscope indicate that the relationship of the myofibrils to the sarcoplasmic reticulum is upset in dermatomyositis. Since a muscle functions adequately only when these structures are correctly apposed to one another, any alteration would doubtless weaken the muscle. How such alterations arise in dermatomyositis is not clear, except that from our evidence the blood-vessels, sarcolemma or subsarcolemmal nuclei show but little change at the outset. In a recent study, the fine structure of blood-vessels in affected muscles was examined under the electron-microscope.[8] Thickening of the endothelium and basement membranes was found, and the endothelial cytoplasm contained autophagic vacuoles. However, these changes were mild and patchy and did not suggest a primary vascular lesion. It also seemed to us as if the relationship between cytoplasm and tonofibril could be disturbed in the epidermal cell, the cytoplasm being reduced and the tonofibrils sometimes standing out sharply as if the cell had been turned into a skeleton. Quite possibly the endothelial cells and fibrocytes in the affected zones are altered, but we are less sure about these findings and their effects on the tissue.

Treatment

Even when the diagnosis of dermatomyositis has been suggested or made, we have observed that cases may easily be undertreated with corticosteroids. Because some cases die and others recover anyway, this hardly warrants therapeutic inactivity. We suspect that certain uncomplicated but severe cases in our series have been saved by timely and intensive corticosteroid treatment and others lost through want of it. Even such changes as the skin sclerosis of dermatomyositis are in our experience reversible with steroid treatment by contrast with scleroderma where they are little influenced. Where the dermatomyositis is complicated (carcinoma, tuberculosis, other concurrent disease, etc.) the treatment and outlook are less hopeful.

ADDENDUM

Recent experience with cytostatic agents has shown that they may largely replace or even displace corticosteroids where the response to treatment is inadequate. We have found cyclophosphamide extremely promising where oral steroid treatment had begun to fail.

SUMMARY

An attempt was made to assess the incidence of dermatomyositis in the Bantu population from certain parts of the Transvaal. The Bantu are probably 10 times more prone to dermatomyositis than the White population. Eleven Bantu and 4 White patients seen recently with dermatomyositis have been reviewed and the clinical features, laboratory studies, prognosis and postmortem findings briefly presented. Some speculations have grown out of the findings, which relate to the triggers for dermatomyositis during the active reproductive period and the relationships between skin and muscle in the pathogenesis.

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