DERMATOMYOSITIS IN THE SOUTH AFRICAN BANTU

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Polymyositis and dermatomyositis, while rare diseases, are well documented in the literature from Europe and North America, as reference to the recent reviews of Bitnum *et al.*[3] and Barwick and Walton[2] will show. Reports from Africa, however, are infrequent. Neither Gelfand[8] nor Trowel[12] recognize this disease syndrome in Central or East Africa. The cases reported from Africa to date are 3 cases with bullous skin lesions by Findlay *et al.*[7] in the South African Bantu, 1 case by Sament and Klugman[11] in a Eurasian in South Africa and 3 cases from West Africa by Barnard *et al.*[1]

There are reports of this condition in the Negro from the United States, e.g. Irgang,[9] Bradley *et al.*,[5] Cook *et al.*[6] and Bitnum.[4] It appears, however, that Negroes do not get this disease as often as other ethnic groups.[4]

Four cases of dermatomyositis seen in South African Bantu females are described in this paper. All the patients were admitted to Jane Furse Memorial Hospital, which is situated in a tribal reserve in the North-Eastern Transvaal. It is of interest to note that the hospital records show that in the period February 1958—June 1963 10 cases classed as acute dermatomyositis (or polymyositis), 1 case of possible past myositis, and 1 case of possible chronic myositis were admitted to this hospital.

SUMMARIES OF CASE HISTORIES

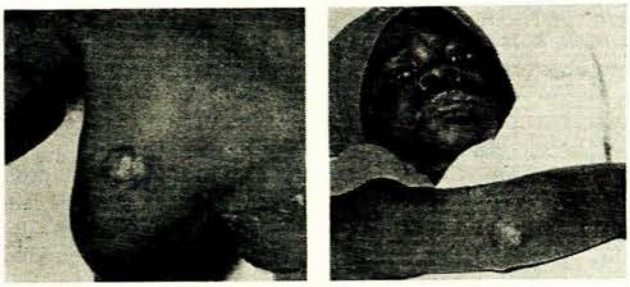
Case 1

M.N., Bantu female aged 30 years, admitted to hospital on 5 August 1960.

Complaint. Swelling of body, skin rash and productive cough of a few weeks' duration.

Past history. Pulmonary tuberculosis treated July 1956 to January 1957. Outpatient chemotherapy with PAS and INH continued for 2 years.

Physical examination. Ill woman, low-grade pyrexia. *Skin*: scaly pigmented macules on extensor surfaces of arms and legs and on the back over scapulae. Generalized oedema with



Figs. 1, 2. The lesions on the elbows and shoulder and puffiness and pigmentation of the face are shown (case 1).

puffiness of the face. Muscles tender. Marked generalized muscle weakness; patient unable to do anything for herself, breathing and swallowing difficult. Reflexes present. *Systemic examination*: No abnormality.

Investigations. X-ray examination of chest: no evidence of active tuberculosis. Urine: no abnormal constituents. Hb. 74%, dimorphic blood picture. ESR 27 mm./hr. Muscle biopsy report: 'Section of this biopsy specimen from the deltoid muscle shows the presence of bundles of striated muscle fibres which in some cases show focal loss of muscle striations. In addition there is fragmentation of muscles with a fairly diffuse infiltrate of the fibres by lymphocytes, histiocytes, a few plasma cells and some eosinophils. The histological features are consistent with dermatomyositis.'

Treatment. Prednisolone and Stenediol followed by ACTH gel. Gradual improvement noted but progress complicated by development of foot-drop and contractures of elbows and by exacerbation of pulmonary tuberculosis necessitating chemotherapy. Corticosteroids discontinued after 38 days' treatment; physiotherapy continued.

Discharge in May 1961—well.

Confinement 1962. No complications.

Re-admitted on 5 August 1963 with similar condition of oedema, muscle tenderness and weakness and similar skin lesions. The face showed a pigmented reticular appearance. Early contractures of elbows noted. Urinary creatinine excretion: 402 mg. in 24 hours. ECG: Flat T-waves. ESR: 40 mm./hr. X-ray chest: normal. Hb. 100% normal blood film. No LE cells.

Treatment: Prednisolone, Durabolin and physiotherapy. INH and Thiacetazone continued.

The patient improved gradually and when last seen on 14 December 1963 was well, apart from residual muscular weakness and wasting.

Case 2

N.M., Bantu female aged 31 years, admitted to hospital on 29 May 1962.

Complaint. Generalized pains and muscular weakness of recent onset.

Physical examination. Generalized muscular weakness, unable to sit up unaided, crawls on hands and knees. Regurgitation of fluids through the nose. Marked muscle tenderness with generalized 'firm' oedema including puffiness of face. Reflexes present. *Systemic examination*: No abnormality. Blood pressure 120/80 mm.Hg. No rash.

Investigation. X-ray examination of chest, routine urine examination, ECG, blood count, Brucella and Widal agglutinations and the Wassermann reaction were all normal. No LE cells were seen. ESR: 13 mm./hr. (Westergren). SGOT: 31 U., SGPT: 26 U., Lactic dehydrogenase: 450 U. Urinary creatinine excretion: 762 mg. in 24 hours. Muscle biopsy report: 'Sections of this specimen of muscle from the extensor muscles of the left forearm showed the presence of the following changes: there are foci of round and mononuclear cell infiltration chiefly in relation to vessels and in some places apparently involving the walls of the vessels. The fibres show a variety of changes. One or two show necrosis, some vacuolation of the sarcoplasm, some basophilia of the sarcoplasm in association with vesicular nuclei and nucleoli. There are also a few foci of haemorrhage and an area of phagocytosis. These changes are in keeping with acute polymyositis.'

Treatment. Treated with ACTH gel and Durabolin with gradual lessening of oedema and muscle tenderness and improvement in muscle power. Discharged after 6 weeks.

Re-admitted on 10 November 1962 with similar condition, responded to prednisone and discharged after one month.

Re-admitted again on 19 July 1963 with condition exactly similar to that on first admission, again improved on prednisone together with physiotherapy. Discharged after 3 months completely recovered.

Case 3

N.S., Bantu female aged 5 years, admitted to hospital on 21 June 1963.

Complaint. Swelling of body for 1 week.

Physical examination. Good nutrition. Generalized oedema. Cries when touched. Unable to sit up. Muscles tender, definite weakness all muscle groups, reflexes present but diminished. *Systemic examination.* No abnormality apart from septic gums.

Investigations. X-ray examination of chest, urine and blood count were normal. Mantoux 1/1,000 negative. Serum pro-

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teins 4·0 G/100 ml. total. Urinary creatinine excretion: 171 mg. in 24 hours. No LE cells seen.

Report on muscle biopsy taken from the right quadriceps muscle: 'Sections of this muscle biopsy show a suggestion of wasting of the muscle fibres with prominence of the subsarcolemmal nuclei. There is a sparse infiltration of the interstitial tissue by plasma cells and lymphocytes. The histological features which are present could be consistent with a polymyositis but are not diagnostic of this condition.'

Treatment. Prednisolone was given, resulting in rapid improvement; discharged after 2 months—clinically well.

Case 4

M.M., Bantu female aged 11 years, admitted to hospital on 23 July 1963.

Complaint. Painful limbs for 4 months. Swelling of body for one week.

Physical examination. Unwell. Low-grade pyrexia. Face puffy. Depigmented and scaling patches extensor surfaces of both arms. Slight oedema of legs and over muscle masses. Muscles of upper limbs tender. Contractures of elbows 90°. Upper limbs weak. Gait unsteady. Unable to sit up unaided. Reflexes all present and equal, diminished. *Systemic examination*: No abnormality.

Investigations. X-ray examination of the chest and elbows, urine, blood count, Brucella agglutination and Mantoux test were all negative. ESR 34 mm./hr. Urinary creatinine excretion: 523 mg. in 24 hours. No LE cells seen. *Muscle biopsy*: 'Sections from this specimen of right deltoid muscle show the presence of striking lesions. A perivascular round-cell infiltrate is present and there is a slight increase in endomysial collagen and fat. In addition there are structural changes in the muscle fibres: vacuolar degeneration, basophilia of sarcoplasm with vesicular nuclei and nucleoli, a loss of sarcoplasm in some parts associated with proliferation of histiocytes suggestive of phagocytosis, and a few ringed fibres. There appears to be an oedematous separation of the muscle fibres as well. On the edge of one fragment there is a moderately large nerve showing destruction of the fibres in the centre. Adjacent to the abnormal nerve is a fairly large area of thin muscle fibres suggesting group atrophy. The histological features, therefore, are in keeping with those of a myositis together with a neural lesion. We have observed group atrophy in association with myositis before (so-called neuro-myositis). The myositis is consistent with that seen in dermatomyositis (polymyositis).'

Treatment. The patient was treated with prednisolone and with physiotherapy. Improvement was gradual but associated with marked residual muscle wasting. This patient subsequently developed a high pyrexia and was found to have a primary tuberculous lung complex which responded to antituberculous chemotherapy, but she had no relapse of acute dermatomyositis.

DISCUSSION

The cases described in this paper all had a strikingly similar picture. The salient clinical features were: an acute onset, with or without low-grade pyrexia, a widespread and progressive muscular weakness with marked muscle tenderness and a diffuse oedema. Skin changes other than oedema were present in 2 patients although the difficulties of recognizing minor degrees of erythema in dark-skinned races will be obvious. Muscle wasting was prominent after the acute phase had passed. The histological features on muscle biopsy were degenerative change in the muscle fibres with perimyseal and perivascular round-cell infiltration.

Treatment with corticosteroids produced a dramatic improvement of the acute symptoms and signs although there was a tendency to relapse, as shown in cases 1 and 2. It may well be that, had it been possible to give a prolonged course of corticosteroids, these relapses would have been prevented.

The classical clinical picture described in dermatomyositis is one of muscular weakness and tenderness, with possible development of contractures, and a variety of skin lesions, commonly erythematous, of a heliotrope hue over 'butterfly' area of face, aggravated by sunlight. Oedema, which may be the dominant feature, is usually present. Residual atrophy of skin over exposed surfaces and possibly calcinosis may occur.[10] The pathological changes seen by Walton and Adams[13] consisted of degeneration of muscle fibres, infiltration of inflammatory cells, extreme variation in size of muscle fibres, regeneration of muscle fibres and fibrosis.

There is still some confusion in the use of the terms polymyositis and dermatomyositis. Walton and Adams[13] and Barwick and Walton[2] give the following classification:

Group I. Polymyositis
- Acute with myoglobinuria
- Subacute or Chronic
  - in childhood
  - in early adult life
  - in middle or late life

Group II. Polymyositis with dominant muscular weakness but with some evidence of an associated collagen disease or dermatomyositis with severe muscular disability and with minimal or transient skin changes.

Group III. Polymyositis complicating severe collagen disease, e.g. rheumatoid arthritis or dermatomyositis with florid skin changes and minor muscular weakness.

Group IV. Polymyositis complicating malignant disease including carcinomatous myopathy and dermatomyositis in patients with malignant disease.

Excluding the group with underlying malignant disease this classification depicts a group of diseases with, at the one end, predominantly muscle lesions and at the other end predominantly skin lesions, with an intermediate group showing both types of lesions. Cases usually described as dermatomyositis would mostly fall into group II in this classification.

Study of our cases will show that all had clinical and pathological features consistent with polymyositis (group I or group II). The cases also bear a strong resemblance to case 1 described by Barnard *et al.*[1] I have preferred to use the term 'dermatomyositis' because, although there were no definite skin changes in 2 of the cases, all appeared to fall into one disease syndrome.

It is surprising that there have been so few reports of dermatomyositis or polymyositis from Africa. It is probable that many diagnosed cases are not reported on and that in many cases the diagnosis is missed. As Walton and Adams[13] point out, although diagnosis is often difficult, a combination of clinical and pathological findings, with the electromyogram and biochemical tests, will define and distinguish polymyositis as a specific syndrome in the majority of cases.

SUMMARY

Attention is drawn to the paucity of reports from Africa on dermatomyositis. In American Negroes the disease also appears to be less common than in those of European descent.

An account is given of 4 South African Bantu female patients from the Transvaal with clinical and histological features of dermatomyositis, all of whom improved on corticosteroids. It seems likely, on the basis of this report, that dermatomyositis is a more common disease in Africa than a study of the literature indicates.

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