Skin changes in sarcoidosis

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Summary

The skin changes which occur in sarcoidosis are erythema nodosum and specific granulomata.

The incidence of erythema nodosum and its frequency in England and Scandinavia is contrasted with its comparative rarity among Negro patients in the United States. The various types of granulomata are described and classified. In contrast with erythema nodosum, granulomata are more common in Negro than in Caucasian patients.

The skin changes which may occur in sarcoidosis are erythema nodosum and sarcoid granulomata, appearing either in normal skin or infiltrating scars. The frequency of these lesions is variable and depends on the sex and race of the patients under observation (Tables 1 and 2). In series from London and Stockholm, the incidence of patients with erythema nodosum roughly equalled the number of patients with specific skin granulomata, whereas among the patients reported from Philadelphia, skin granulomata were fairly frequent while erythema nodosum was uncommon both in Negroes and Caucasians.

Erythema nodosum

Three series have been reported from London of the frequency of erythema nodosum in sarcoidosis (Table 1). There was a close correlation between Scadding's patients (1967) of whom 11% were affected and those of Smellie & Hoyle (1960), of whom 13% were affected. A third of James' (1959) patients had erythema nodosum but this may be an artificially high figure due to referral of cases to this

author because of his well-known interest in the disease.

In Stockholm the condition is probably more frequent than in London, an incidence of 25% being reported by Löfgren & Stavenow (1961). In contrast, of the 211 patients seen by Sones & Israel in Philadelphia (1960), only 2.9% had erythema nodosum; this group comprised 184 Negroes, of whom only four women (2%) had erythema nodosum and twenty-seven Caucasians of whom two women (7%) were affected.

From clinical experience erythema nodosum seems much more common in women than men but this impression is biased due to the more frequent occurrence of sarcoidosis in women than men. When the sex incidence is compared on a percentage basis, the incidence of females to males was 3:1 (Scadding, 1967), 2:1 (James, 1959) and 5:1 (Löfgren & Stavenow, 1961).

Although in adults erythema nodosum may be fairly frequently associated with sarcoidosis, this was not its most common aetiology in the seventy patients attending dermatological out-patients in Buckinghamshire and reviewed by Vesey & Wilkinson (1959). They concluded that of this group 46% were of proven or presumed streptococcal origin, 36% were associated with sarcoidosis, 6% were tuberculous and the rest were of mixed or unknown origin. These figures agree with those of a small group of nine patients with erythema nodosum seen and investigated in the dermatology department of this hospital during the past 9 years. The aetiology was thought to be streptococcal in three patients, associated with sarcoidosis in another three and of

TABLE 1. Incidence of erythema nodosum in sarcoidosis in London, Stockholm and Philadelphia

Place	Series	Number of patients	Erythema nodosum	F : M
London	Scadding (1967)	275	11%	3 : 1
	James (1959)	200	31%	2 : 1
	Smellie & Hoyle (1960)	125	13%	
Stockholm	Löfgren & Stavenow (1961)	132	25%	5 : 1
Philadelphia	Sones & Israel (1960)	211	2.9%	

TABLE 2. Incidence of skin granulomas in sarcoidosis in London, Stockholm and Philadelphia

Place	Series	Number of patients	Skin granulomas
London	Scadding (1967)	275	15.3%
	James (1959)	200	16.5%
	Smellie & Hoyle (1960)	125	13.6%
Stockholm	Löfgren & Stavenow (1961)	132	32%
Philadelphia	Sones & Israel (1960)	211	26.5%
			Negroes = 29%
			Caucasians = 11%

the remaining patients, psittacosis was the cause in one, it followed sulphonamide therapy in another and in the last patient the cause was not determined.

Granulomatous lesions

The frequency of granulomatous lesions appearing in normal skin or scars is also variable. The three series from London (Table 2) are remarkably similar varying from 16.5% (James, 1959) to 13.6% (Smellie & Hoyle, 1960). There is a higher incidence of lesions in Stockholm, 32% being observed by Löfgren & Stavenow (1961), these were mainly infiltrations of old scars by small granulomas. American Negroes have a definite tendency to develop skin sarcoidosis, 29% of those reported by Sones & Israel (1960) being affected; by comparison only 11% of the Caucasians in Philadelphia had these lesions (Table 2).

Classification of cutaneous sarcoidosis has to be based on the clinical appearance of the lesions. As the granulomas may invade the skin at any level, may be small or large and localized or diffuse, a comprehensive morphological classification has of necessity several sub-groups. The various types of lesions which may occur are listed in Table 3, and any patient may have more than one type of lesion.

Plaques

Lupus pernio is the most characteristic of all the morphological patterns. Bluish purple thickenings occur on the nose, cheeks, ear-lobes, fingers and over the knees; the changes can be striking and

disfiguring and may be the presenting sign of the disease. Similar types of plaques may occur on the trunk and limbs. When the scalp is involved it causes scarring alopecia (Bluefarb, 1955; Scadding, 1967). These lesions are very persistent and may last up to 36 years (Scadding, 1967); they may be associated with progressive fibrosis of the lungs, bone cysts and in some patients uveitis.

A patient Mrs A, 47 years, has had disfiguring lupus pernio and plaques on her body for 30 years. She has hilar lymphadenopathy but no other systemic abnormality.

Nodules

Large and small nodules may occur anywhere on the body and in any level of the skin or subcutaneous tissue. The larger ones tend to persist and may be associated with pulmonary fibrosis, the smaller ones tend to resolve as do also the lung changes (Scadding, 1967). Two Negro patients with cutaneous nodules were described by Marten & Warner (1967). They are similar to the patient described below:

Mrs C., 36 years, a Jamaican was found, in November 1968, to have bilateral hilar gland enlargement and reticulation in both lung fields. Her Kveim test was positive and a gland removed at mediastinoscopy was replaced by epithelioid granuloma. In May 1969 she developed nodules on her arms and thighs, which come and go spontaneously, initially there was no surface change but recently there has been slight scaling over some of the lesions. In September she suddenly developed bilateral uveitis. The histology of an early cutaneous nodule removed in May showed an encapsulated subcutaneous inflammatory granuloma consisting of polymorphs, leucocytes and a few eosinophils. A second nodule removed in July showed a rather diffuse epithelioid granuloma with poor follicle formation and a further nodule biopsied in September showed a similar epithelioid granuloma with inflammatory reaction.

Papules

A sheeted eruption of small pin-head sized (lichenoid) papules may occur on the trunk and

TABLE 3. Classification of cutaneous granulomas in sarcoidosis

Skin	1. Plaques	Lupus pernio Scattered on body Annular
	2. Nodules	Dermal—small and large Subcutaneous
	3. Papules	
	4. Scars	
	5. Erythroderma	
	6. Ulcers	
Mucosae		
Nails		

Plate 2

Explanation to Plate 2

2a. Jamaican female of 26 years with small dermal nodules on the limbs; associated hilar and mediastinal lymphadenopathy.

2c. A male aged 22 years with erythrodermic sarcoidosis and associated involvement of lymph nodes. Chest X-ray was normal.

2b. West Indian woman of 37 years with papular lesions on the face, associated involvement of lymph nodes and nasal bones. Lung fields clear on X-ray.

2d. West Indian male of 37 years with destruction of nails and cyst formation in the underlying phalanges, also cutaneous granuloma of upper lip. Chest X-ray normal.

limbs. The lesions erupt suddenly and may appear in crops.

In Negro patients a very characteristic pattern is the eruption of discrete papules or small papulonodules along the eyelids, around the nasal orifices, over the pinnae and scattered on the forehead, face and over the trunk (Longcope & Freiman, 1952). There may be few or many lesions.

Infiltration of scars

Infiltration of old or new cutaneous scars is well known and was the commonest form of skin sarcoidosis reported from Stockholm (Lofgren & Stavenow, 1961).

Erythroderma

Generalized erythroderma due to widespread infiltration of the skin by granulomata does occur but is very rare. A man of 22 years with this type of eruption was described by Wigley & Musso (1951); the eruption persisted for 5 years and then faded after a course of calciferol.

Ulcers

Ulceration of the skin is very uncommon. It has been described in Negro patients by Irang (1965) and in this country by Simpson (1963) in a very unusual patient who had both erythroderma and cutaneous ulcers.

Mucosae

Nasal obstruction and widening of the bridge of the nose due to involvement of the nasal bones and mucosae were the presenting signs in eleven patients described by Porter, Stevenson & Munro-Black (1970).

Nails

Thickening and distortion of nails is sometimes seen and is associated with changes of the underlying terminal phalynx (Scadding, 1967).

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References

- BLUEFARB, S.M., SZYMANSKI, F.J. & ROSTENBERG, A. (1955) Sarcoidosis as a cause of patchy alopecia. *Archives of Dermatology*, **71**, 602.
- IRANG, S. (1955) Ulcerative cutaneous lesions in sarcoidosis. *British Journal of Dermatology*, **67**, 255.
- JAMES, D.G. (1959) Dermatological aspects of sarcoidosis. *Quarterly Journal of Medicine*, **28**, 109.
- LÖFGREN, S. & STAVENOW, S. (1961) Course and prognosis of sarcoidosis: Stockholm. *American Review of Respiratory Diseases*, **84**, 71.
- LONGCOPE, W.T. & FREIMAN, D.G. (1952) A study of sarcoidosis. *Medicine*, **31**, 1.
- MARTEN, R.H. & WARNER, J. (1967) Subcutaneous nodular sarcoid. *Transactions of St John's Hospital Dermatological Society*, **53**, 160.
- PORTER, D.I., STEVENSON, C.J. & MUNRO-BLACK, J.M. (1970) (to be published).
- SCADDING, J.G. (1967) *Sarcoidosis*. Eyre & Spottiswoode, London.
- SIMPSON, J.R. (1963) Sarcoidosis with erythrodermia and ulceration. *British Journal of Dermatology*, **75**, 193.
- SMELLIE, H. & HOYLE, C. (1960) The natural history of pulmonary sarcoidosis. *Quarterly Journal of Medicine*, **29**, 539.
- SONES, M. & ISRAEL, H.L. (1960) Course and prognosis of sarcoidosis. *American Journal of Medicine*, **29**, 84.
- VESEY, C.M.R. & WILKINSON, D.S. (1959) Erythema Nodosum. *British Journal of Dermatology*, **71**, 139.
- WIGLEY, J.E.M. & MUSSO, L.A. (1951) A case of sarcoidosis with erythrodermic lesions. Treatment with calciferol. *British Journal of Dermatology*, **63**, 398.