

Correlation between Cutaneous Sarcoidosis and Systemic Sarcoidosis

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Twenty patients with cutaneous sarcoidosis and 21 patients carrying isolated skin sarcoids were studied. (We use the term 'sarcoid' to emphasize that exclusively skin was altered.) Both groups were compared by clinical and histological patterns and certain data concerning the state of the mononuclear phagocyte system (MPS). It was found that skin lesions in sarcoids and sarcoidosis do not differ regarding either in clinical or histological manifestations. The changes in the functional activity of monocytes and macrophages were the same. The data obtained allow us to suggest that sarcoids should be regarded as a systemic disease connected with changes in the MPS reaction. *Key word: Mononuclear phagocyte system.*

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Skin lesions in sarcoidosis are the key to the discovery of damage in other organs (1). Lesions in several organs can be regarded as sarcoidosis, while those observed solely in the skin are called sarcoids. Our aim was to study the relationship between these two types.

PATIENTS AND METHODS

We investigated 4 men and 16 women with cutaneous sarcoidosis and 4 men and 17 women with isolated skinsarcoids. Diagnosis was based on X-ray examination, tomography of chest organs, roentgenographic images of bones in hands and feet, and histology of skin lesions. An ophthalmologist was engaged for consultations.

In 15 patients with sarcoidosis and in 13 with sarcoids the functional activity of monocytes in the integral test was measured using nitroblue tetrazolium (NBT-test). One hundred monocytes were counted in each preparation, the cells being distributed in five grades of activity: zero grade denoting functionally inactive cells, 1st-2nd grades denoting evidence of metabolic activity, while cells with 3rd-4th grade activity are phagocytosing. The percentage of functionally active cells (PFAC), reflect the activity of dehydrogenases, was estimated using these grades as well as the coefficient of functional activity (CFA). The percentage of phagocytosing monocytes (PPM) reflects the intensity of phagocytosis and its completeness with respect to the activity of oxidases in phagosomes, and the phagocytosis activity coefficient (CAP).

Table I. *Clinical characteristics of patient with sarcoidosis and sarcoids*

	Sarcoidosis	Sarcoid
Papular lesions	16	12
Macular lesions	2	3
Lupus pernio	1	2
Subcutaneous lesions	-	3
Scar infiltration	1	-
Ulcerative lesions	-	1

Ten patients with sarcoidosis and 8 with sarcoids underwent the isotope test, using ^{99m}Tc -phosphon in the gamma-counter MB-91009101A (Hungary). An isotope preparation with the activity 400 MBq was injected intravenously and the bones were subjected to scintigraphy.

RESULTS

The data presented in Table I show that the clinical picture of skin lesions is the same in patients with sarcoids as for those with sarcoidosis. No correlation was observed between the spread of the skin lesions and the damage to internal organs and bones. Alterations in lungs and bilateral hilar lymphadenopathy preceded the skin lesions in 12 patients, while in 2 cases, pulmonary alterations developed later. Uveitis was seen in 3 patients and changes in hands and feet in 9.

Histology of skin lesions did not reveal any peculiarities in the structure of granulomas in sarcoids or sarcoidosis, but did visualize focal collections of epithelioid cells surrounded by a narrow rim of lymphocytes.

Our data on the MPS (Table II) show that the metabolic and phagocytotic activity of monocytes is sharply quenched in both sarcoid and sarcoidosis, the degree of inhibition being independent of the organ affected, the number of organs involved in the process, and the duration of the disease. Isotope monitoring of the MPS demonstrated a non-typical distribution of the label in long tubular bones and skull, in the case of both sarcoid and sarcoidosis, which is regarded as evidence of the increased activity of macrophages.

DISCUSSION

The current definition of sarcoidosis implies that the disease can be diagnosed only when more than one organ or tissue is affected by epithelioid-cell tubercles without caseation (2,3,4). Therefore, when epithelioid-cell tubercles are detected solely in the skin (without lymphocytes), one should not diagnose sarcoidosis. Rather, it is reasonable to regard the

Table II. *Functional activity of monocytes in patients with sarcoidosis and skin sarcoids ($\bar{x} \pm m\bar{x}$)*

	Proportion of active cells (%)	Proportion of phagocytosing cells (%)
1. Control group (n = 42)	76 \pm 3.2	27.3 \pm 2.1
2. Patients with sarcoidosis (n = 15)	41.1 \pm 4.1*	9.7 \pm 2.6*
3. Patients with skin sarcoids (n = 13)	40.0 \pm 4.0*	8.8 \pm 3.0*

* Significantly different from the controls ($p < 0.01$).

condition as a sarcoid reaction (5) specific to an injury (6). Moreover, Callen (7) and Veien et al. (8) believe it to be impossible to differentiate between clinical and histological skin changes in isolated epithelioid-cell tubercle involvement when other organs are involved in the process. It is also a common opinion that there is an opportunity for isolated skin sarcoidosis (8).

The data presented allow us to suggest that skin sarcoid should be regarded as a systemic disease as stipulated by the MPS reaction. Lesions in organs, specifically the skin, are the 'top of the iceberg', with MPS changes being 'beneath the surface'. At the same time it must be taken into account that skins arcoidosis can exist in isolation for unlimited periods of time.

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