MEDICAL GRAND ROUNDS
PARKLAND MEMORIAL HOSPITAL
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"THE ENIGMA OF SARCOIDOSIS"

Second Conference on Sarcoidosis, National Research Council, October 1956:

"Sarcoidosis is a systemic disease, or group of diseases, of undetermined etiology and pathogenesis. Histologically, it is marked by the presence of epithelioid-cell tubercles, showing little or no necrosis. Varying types of inclusions in giant cells may be present but are not pathognomonic. A similar histological picture may be found in certain other diseases, especially in infectious granulomas and in beryllium poisoning. Clinically, the disease most frequently involves lymph nodes, lungs, skin, eyes, liver, spleen and phalangeal bones. The course is usually chronic and constitutional symptoms vary markedly. More specific symptoms, when present, relate to the tissues and organs involved.

The intracutaneous tuberculin test is frequently negative, but a positive test does not controvert the diagnosis. Hyper-(gamma)-globulinemia and leucopenia are common and hypercalcemia, hypercalcinuria, elevated alkaline phosphatase and eosinophilia are variable, but sometimes significant features of Sarcoidosis.

The diagnosis of Sarcoidosis is based upon the above clinical features associated with a compatible histological picture, provided beryllium poisoning and infectious processes of known etiology can be excluded. Spontaneous clinical recovery, with or without recognizable fibrosis, may result or Sarcoidosis may persist for years with varying functional alteration of the

tissues and organs involved, or the disease may follow a progressive course, ending fatally."

Etiology:

"In the present state of our knowledge, it would seem wiser to recognize clearly the complexities that confront etiological studies and methodological limits of our present working tools than to insist on any one etiological hypothesis, including the agnostic one that claims that the etiology is unknown." (Pinner 1946).

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The above statements bring our lack of knowledge clearly into focus. Because Sarcoidosis is a diffuse granulomatous process, its clinical expressions involve many different medical disciplines, often with distortion of the true course and nature of the disease.

Primary Syndrome:

Bilateral hilar adenopathy, fever, cough, uveitis, arthralgias and/or arthritis, splenomegaly and erythema nodosum. Onset insidious in young adults, predominantly caucasian. Most severely ill are those with erythema nodosum and arthritis. Course: Symptoms fade in a few days to a few weeks, nodes regress in 3-6 months and only a very small percentage are believed to develop progressive Sarcoidosis.

Differential Diagnosis: Drug hypersensitivity, streptococcal infection, other granulomatous diseases (esp. histoplasmosis and coccidiodomycosis), Sjögren's syndrome and lymphoma.

Diagnosis of Early Sarcoidosis:

1. Erythema nodosum and/or arthritis
2. Bilateral hilar adenopathy
3. Uveitis
4. Biopsy confirmation

Course of Early Sarcoidosis:

(A) Acute phase:

Fever, malaise - short duration

Erythema nodosum and arthritis fade in 1-3 months

BHA disappears in 12 months, usually in first 3-6 months

(B) Subacute or Chronic phase:

Only a small percentage enter this phase; this group then constitutes the patients with remissions and exacerbations or with progressive disease.

Proposed Clinical Sequence for Sarcoidosis:

1. Primary Syndrome
2. Clinical evidence of disseminated disease
3. Progressive organ functional involvement
4. Fibrosis

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References:

1. Sjogren's Syndrome; Weissmann, G. - American Journal Medicine 24:475-481, 1958

(References continued)

2. Primary Syndrome:

(a) James, D. J. Postgraduate Medical Journal 34:240, 1958

(b) Fløystrup. Acta Tuberculosa Scand. Supp. XLVII, June 1959

(c) Smellie and Hoyle - Lancet 273: 66-70, 1957

(d) Lofgren. Acta Medica Scand. 145:424, 1953

(e) Turiaf, J. and Brun, J. La Sarcoidose Endothoracique de Besnier-Boeck-Schaumann, p. 115, 1955 (Paris, France).

General:

Longcope, W. T. et al. Medicine 31:1-132, 1952

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Charles LeMaistre, M.D.
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