Sarcoidosis in identical twins [1]

by

Howard E. Allen, M. D.

Portland, Oregon

Sarcoidosis has been the subject of more than 300 medical articles in the past seventy-five years. Since these are so readily available, no attempt will be made to review the academic information on the subject contained therein (1, 2, 3, 4, 5, 6, 7, 8).

The cause of this granulomatous entity remains undetermined despite attempts in the past to make it one with that of tuberculosis. Since its description by Boeck in 1899 and the subsequent recognition of its protean character and frequent wide distribution through practically all of the body tissues, it has become a disease to be borne in mind whenever diagnosis is obscure.

Its erratic and unpredictable course, with long remissions and even occasional complete clinical healing, is well known. Therapy has included most known agents, but it seems likely that nothing specific will be found until the cause of the disease is determined. Recent reports stress the regression of new lesions, particularly those of the lungs and the eyes, after administration of ACTH and cortisone. However, the physiologic mechanisms involved remain obscure. Further evaluation can be made only when more data have accumulated.

No one seriously believes that sarcoidosis, or a predisposition to the disease, is inherited. Nonetheless, there are accounts of families in which more than one member has been affected by the disease, often at the same time. Indeed, Sherer and Kelley (10) in 1949, described the simultaneous occurrence of sarcoidosis, with almost identical signs and symptoms, in two Negro men 22 years of age who were identical twins. Gilg (3), in 1952, reported a similar example of the disease in two 39yearold women who were identical twins and who each had plaquelike lesions on the forehead. Yet a third instance of sarcoidosis in enzygotic twins, with involvement chiefly of the lymph nodes and the parotid glands, was reported by Rogers and Netherton in 1954 (9).

Such unusual developments might be ascribed to constitutional defects or to contact infection. But they could equally well be explained as arising from some external ingested or inhaled infecting agents to which the twins were concurrently exposed.

Because of the exceptional rarity with which sarcoidosis has been observed in identical twins, there appear below the case histories of another pair. The disease occurred almost simultaneously in both, and to date has run a similar clinical course.

[1] Previously published in the Portland Clinic Bulletin, Volume 9, No. 1, June 1955, Portland, Oregon.

Ricevuto il 5. IV. 1956.

Case Reports

Case 1. Mr. N. C. D., a court reporter, aged 24, was first seen at The Portland Clinic on November 9, 1953 because of epistaxis. Only a large dilated vein in the floor of the left nostril was found.

On December 12, 1953 he clipped the end from a wire recording spool and felt something strike his left eye. For the next three or four days he noted slight blurring of vision in

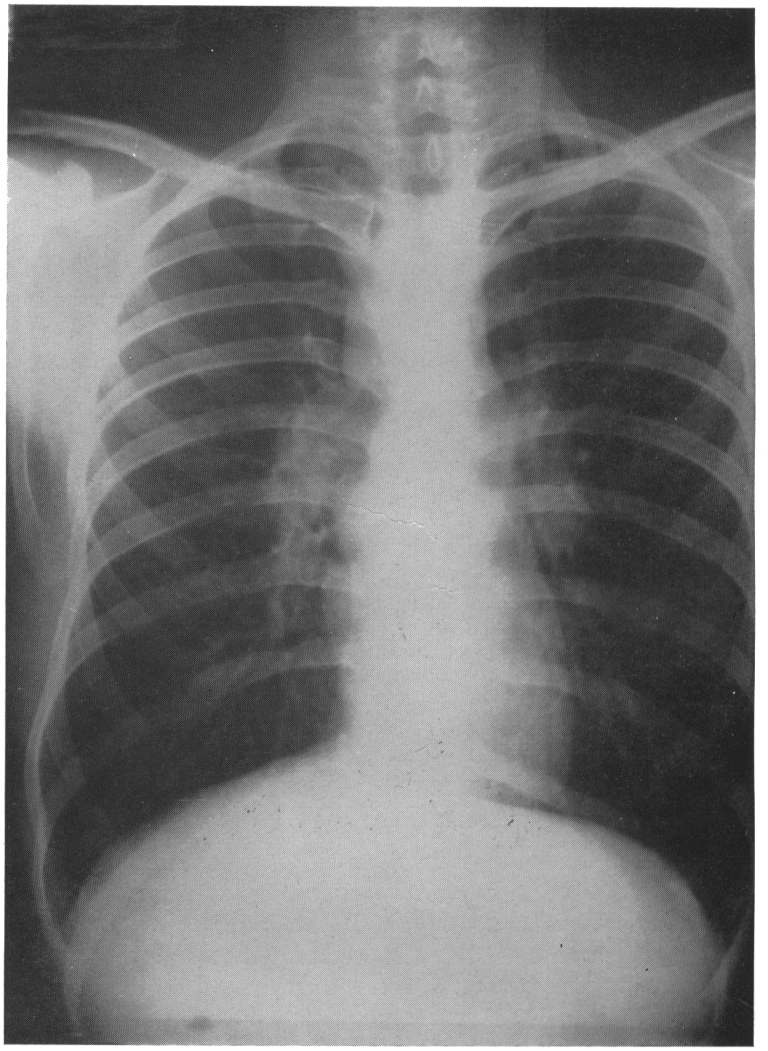

Fig. 1a

https://doi.org/10.1017/S1120962300020072 Published online by Cambridge University Press

this eye. During December he also noted a decline in his weight from 128 to 114 pounds.

Past medical history and systemic review were not remarkable.

On December 18 he was seen by Dr. Robert E. Fischer who noted that the central portion of the corneal epithelium of the left eye was plastered with punctate keratoses resembling mutton fat. Several large Koeppe nodules were present at the pupillary margins. The right eye was normal. Treatment consisted of cortisone drops in the eye three times daily.

General physical examination revealed an underweight, asthenic man measuring 68½

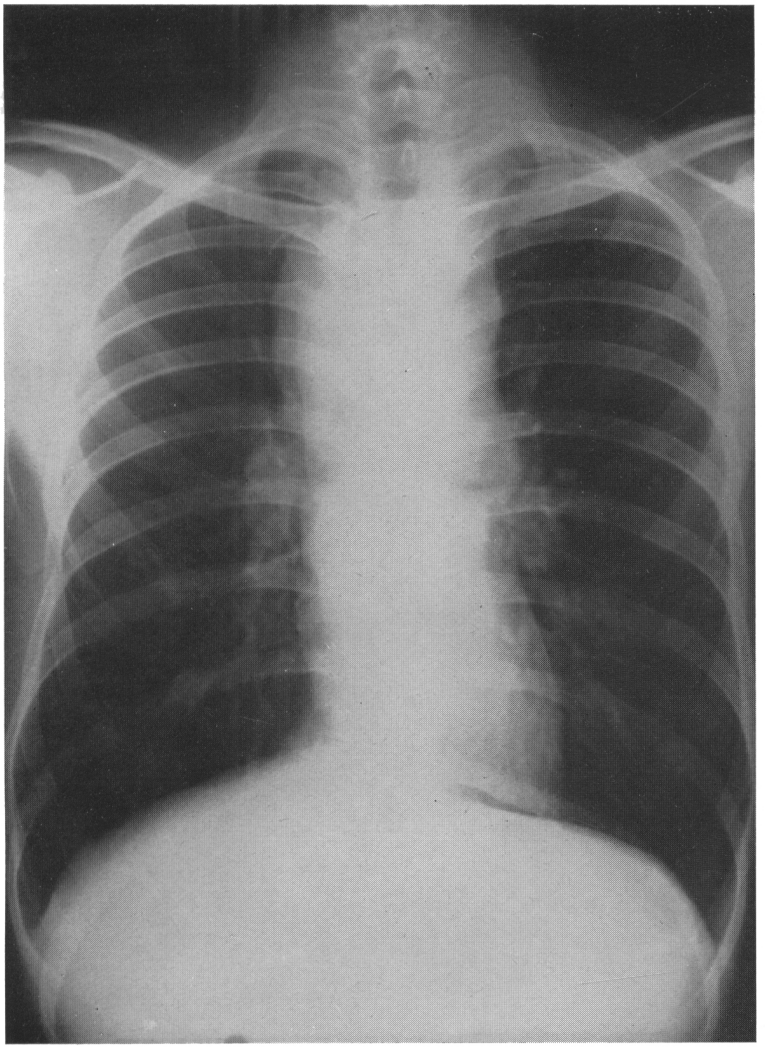

Fig. 1b

https://doi.org/10.1017/S1120962300020072 Published online by Cambridge University Press

inches in height and weighing 118 pounds without clothing. Blood pressure was 110/70 and pulse rate was 84 beats per minute. In all other respects the physical examination was within normal limits. No adenopathy, splenomegaly or hepatomegaly was noted.

Complete blood count, sedimentation rate, Kahn and Kline tests, as well as urinalysis, were negative. A heterophil agglutination test was positive in 1 to 8 dilution.

Roentgenograms of the chest showed definite lobulated masses in both hilar regions and in the right peritracheal region, just below the anterior end of the right clavicle. These

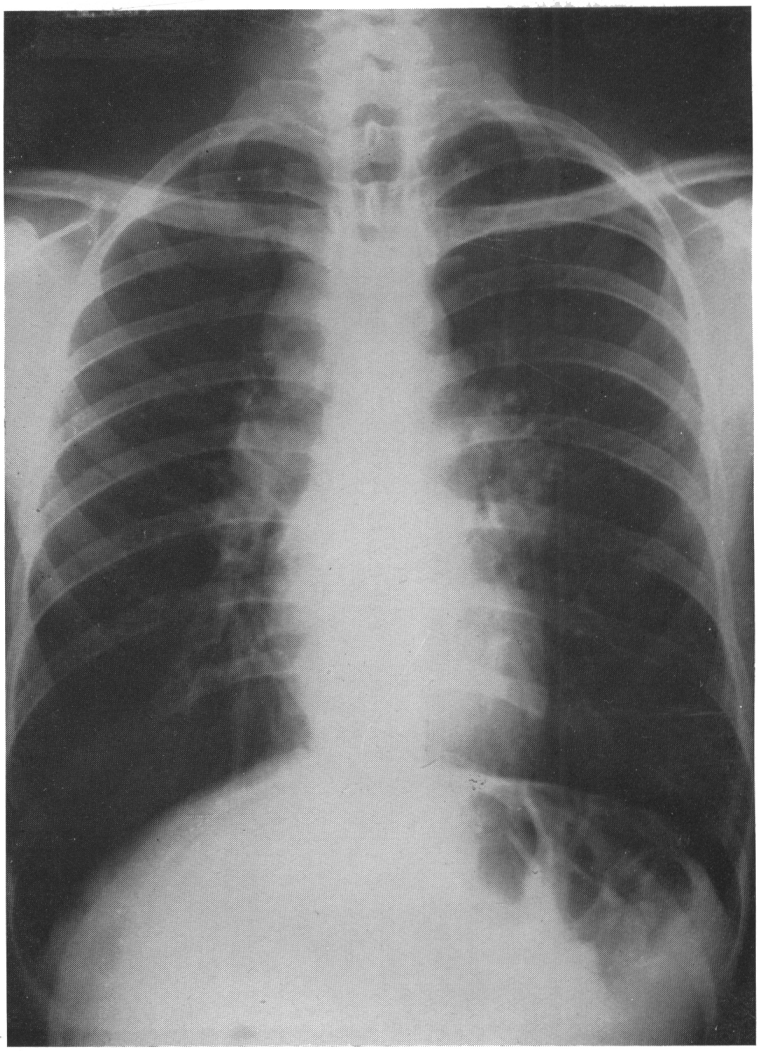

Fig. 2

https://doi.org/10.1017/S1120962300020072 Published online by Cambridge University Press

findings were unchanged in April 1954. At this time a tuberculin test, using second strength P. P. D., was done. It was within normal limits.

During the last two weeks of July the patient was much troubled with nausea and vomiting. Thereafter he gained in weight to 130 pounds. Re-examination of the eyes on October 29 showed only four crenated punctate keratoses on the left cornea. The chest lesions, followed by fluoroscopic examination, underwent no change.

In February 1955 cervical adenopathy was noted for the first time. Biopsy of a lymph node, on February 12, 1955, disclosed the typical picture of sarcoidosis. During the past year roentgenograms of the chest have shown slight progression in the hilar adenopathy (fig. 1a, 1b).

Case 2. Mr. N. J. D., the identical twin of the patient in Case 1, was seen on January 21, 1954 because his eyes, especially the left, had shown some tendency to be bloodshot during the preceding six months.

On examination both corneas presented many punctate keratoses which had the appearance of mutton fat. No Koeppe nodules were noted. A diagnosis of granulomatous uveitis was made.

Roentgenogram of the chest showed a dense, lobulated mass measuring up to 4 cm. in diameter and extending to both the peritracheal and the peribroncheal regions (fig. 2). Peripheral lung fields were clear. The cardiac shadow was within normal limits.

Laboratory studies included blood count, urinalysis, Kahn and Kline tests, and a brucella agglutination determination. All results were within normal limits.

Biopsy of a scalene node, by Dr. J. Karl Poppe on May 8, 1954, was reported as showing typical sarcoidosis.

Summary

The simultaneous occurrence of sarcoidosis in two 24yearold white men who where enzygotic twins is reported. Manifestations of the disease as well as its clinical course were identical in both twins.

References

1. FREIMAN, D. G.: Sarcoidosis, New England J. Med. *239*: 664-671 (Oct. 28) 1948; 709-716 (Nov. 4) 1948; 743-749 (Nov. 11) 1948.
2. GENDEL, B. R., YOUNG, J. M., and GREINER, D. J.: Sarcoidosis. A review with 24 additional cases, Am. J. Med. *12*: 205-218 (Feb.) 1952.
3. GILG, I.: Boeck's sarcoid in identical twins, Acta dermat.-venereol. (supp. 29) *32*: 108-115, 1952.
4. LONGCOPE, W. T., and FREIMAN, D. G.: A study of sarcoidosis based on a combined investigation of 160 cases including 30 autopsies from The Johns Hopkins Hospital and Massachussetts General Hospital, Medicine, *31*: 1-132 (Feb.) 1952.
5. MICHAEL M. Jr., COLE, R. M., BESSON, P. B. and OLSON, B. J.: Sarcoidosis. Preliminary report on a study of 350 cases with special reference to epidemiology, Am. Rev. Tuberc. *62*: 403-407 (Oct.) 1950.
6. REISNER, D.: Boeck's sarcoid and systemic sarcoidosis (Besnier-Boeck-Schaumann disease). A study of 35 cases, Am. Rev. Tuberc. *49*: 437-462 (May) 1944.
7. RICKER, W., and CLARK, M.: Sarcoidosis. A clinicopathologic review of 300 cases, including 22 autopsies, Am. J. Clin. Path. *19*: 725-749 (Aug.) 1949.
8. RILEY, E. A.: Boeck's sarcoid. A review based upon a clinical study of 52 cases, Am. Rev. Tuberc. *62*: 231-285 (Sept.) 1950.
9. ROGERS, F. J., and NETHERTON, E. W.: Sarcoidosis in identical twins, J.A.M.A. *155*: 974-976 (July 10) 1954.
10. SHERER, J. F., and KELLEY, R. T.: Sarcoidosis in identical twins, New England J. Med. *240*: 328-330 (March 3) 1949.

RIASSUNTO

Viene descritta una coppia di gemelli monozigotici di razza bianca aventi 24 anni i quali presentano simultaneamente sarcoidosi. Le manifestazioni della malattia come pure il suo decorso clinico furono identici in entrambi i gemelli.