

## THE POSSIBLE ROLE OF LEPROSY IN THE ETIOLOGY OF THE BESNIER-BOECK SARCOID AND SCHAUMANN'S SYNDROME

BY PROFESSOR JOHN REENSTIERNA, M.D.

*University of Upsala, Sweden*

In the history of the sarcoids two periods are to be strictly differentiated: (1) The *dermatological* one, in which came Besnier's description of lupus pernio and Boeck's description of the multiple benign sarcoids of the skin, and (2) the *syndrome* one, which dates from Schaumann's description of the autonomous syndrome (or disease) lymphogranulomatosis benigna, of which as Schaumann has shown lupus pernio, Boeck's nodular sarcoids, and some of his papular sarcoids are occasional localizations in the skin. In this second period also came Schaumann's description of the erythrodermic manifestations of lymphogranulomatosis benigna.

During the dermatological period the etiological study held good, quite as a matter of course, for each one of the two first-mentioned skin affections, whose genetic identity was then not known. Besnier himself regarded his lupus pernio as lupus erythematosus, whereas later investigators as a rule described it as a variety of lupus vulgaris (Lewandowsky, Grouven, and others), or as a tuberculide (Lenglet, Darier, Jadassohn, and others). Boeck in the beginning suspected an autotoxic origin, later on an infectious-toxic one, possibly tuberculous, for his cutaneous sarcoids. In the standard textbooks the cutaneous sarcoids were as a rule classified among the tuberculides; they were looked upon, together with for example erythema induratum, as skin reactions to bacillary emboli from some internal tuberculoid focus in an individual in a condition of allergy.

The clinical, and often also the histological, analogies with the cutaneous sarcoids which tertiary syphilides, leprous eruptions, etc., were sometimes found capable of displaying made some authors begin to ask themselves if the sarcoids were perhaps simply nothing else but morphologically peculiar skin eruptions of different origins, i.e., not originating from tuberculosis solely, but also from syphilis, leprosy, etc. A convinced representative of this conception was Pautrier who, in 1914, described cases of tertiary syphilides whose characters he considered necessitated the diagnosis of sarcoid.

Even leprosy could, it was found, in some cases display a striking similarity to the sarcoids, so that diagnostic difficulties could occur. Thus Lewandowsky, whose monograph "Die Tuberkulose der Haut" appeared at the end of the dermatological period, wrote regarding Boeck's sarcoids that the microscopical differential diagnosis "besonders von tuberculoider Lepra in Stiche lassen kann." Boeck himself gave a frequently quoted example of the correctness of this, when he once diagnosed sections of tuberculoid leprosy as a cutaneous sarcoid.

Thus during the dermatological period of the sarcoids it was, for clinical and histological reasons, quite natural in certain quarters to look upon the sarcoids as dermatoses of varying nature. The fact that they were considered as possibly being *inter alia* of leprous origin was not so very surprising, since as has been said leprous skin lesions were found whose microscopic characters could not be differentiated from the "sarcoid" structure.

Since Schaumann described the syndrome of lymphogranulomatosis benigna, however, the problem of the possible leprous nature of the sarcoids must be set up quite differently. The question now is whether or not leprosy is able to cause this syndrome, can become generalized in the lymphoglandular system—in the tonsils, the bone-marrow, the spleen and liver, and the lungs, and not only in some gland or other—and can produce in all these tissues identical changes in the form of circumscribed tuberculoid foci without necrosis.

With regard to the question of whether or not the micro-organism of leprosy is able to cause the systemic reactions which form and compose the said lymphogranulomatous syndrome, Rabello's studies (1) seem to me to leave a great deal to be desired. His histological examinations have all been made on skin lesions; in none of his cases have the lymphatic glands or the tonsils been examined, which in view of the predilection of lymphogranulomatosis benigna for the lymphatic system would have been of the utmost importance. As for the skin lesions in Rabello's cases, they displayed disturbance of sensibility. It should be noted on the one hand that such disturbances exist in cases of leprosy in which there is no evidence of the occurrence of the disease in the "systemic" form, which Rabello considers to exist, while on the other hand they do not exist in the skin manifestations of lymphogranulomatosis benigna.

In Rabello's cases radiological examinations have been made

of the lungs and of the bones of the hand and foot, and the author finds such consonance between the changes demonstrated and those in cases of lymphogranulomatosis benigna that he considers the similarity to support his conception that that disease may be of leprous nature.

The fact of the matter is, however, that the radiological picture of the lungs in lymphogranulomatosis benigna is not pathognomonic, and does not permit of any safe diagnostic conclusions unless the case is known otherwise. Thus, for example, chronic miliary tuberculosis can give radiographs which cannot be differentiated from those in lymphogranulomatosis benigna. That leprosy is able to cause similar radiological lung pictures cannot, a priori, be altogether negatived, but as far as I know it has not been proved. Be this as it may, in view of the fact that the radiological examination is an indirect method it would, of course, have been interesting to know the anatomical substratum of the radiological spots and streaks in the lungs which occurred in Rabello's cases. Only autopsy examinations would give a demonstration as to whether or not they are of the same anatomo-pathological nature as the changes which were demonstrated in the skin.

The same remarks hold good as regards the radiographic bone lesions. As Murdock and Hutter (2) have shown, nodular leprosy can cause peculiar changes of the bone; these display radiologically a striking consonance with those in lympho-granulomatosis benigna. The bone changes demonstrated by Rabello coincide perfectly with those of the "osteoporous" type which Schaumann has described in that disease, but from the viewpoint of his hypothesis it would have been valuable had Rabello made histological examinations to see if those changes were accompanied by the same conditions which Schaumann has proved to occur in lymphogranulomatosis benigna. He found that the changes in the bone result from localization of the characteristic granulation tissue in the bone-marrow, and that the medullary lesions are much more extensive than a radiological examination would lead one to believe.

Recently Schaumann has reported the autopsy findings in four cases of lymphogranulomatosis benigna. In two of these cases there was no tuberculous complication; in the other two cases, on the other hand, there was such a complication. In all four cases the disease was generalized in the manner which he had described from the beginning; i.e., it had localizations—with the typical granulomatous tissue or remnants of it—in the whole of the lymphatic gland system and in the bone-marrow, and also in the spleen, liver and lungs. In

view of this dissemination, so characteristic of the disease, to organs which constitute the main localizations of the reticulo-endothelial system, the facts communicated by Rabello seem to me insufficient for basing upon them the answer to the question of whether or not leprosy can cause Schaumann's syndrome. Autopsies with thorough examinations, both histological and bacteriological, of the hemato-poietic apparatus and other organs would be necessary to enable one to answer this question.

#### SUMMARY

1. Leprosy may manifest itself in the form of skin affections which histologically coincide with Besnier's lupus pernio, Boeck's cutaneous sarcoids and Schaumann's erythrodermia.

2. Leprosy is able to cause in the bones, and perhaps also in the lungs, changes which radiologically are strikingly like those that have been observed in connection with those skin affections.

3. It has not been shown or proved that the micro-organism of leprosy can cause the systemic reactions which together constitute Schaumann's syndrome.

4. In order to show that such might be the case it would be necessary to make specially directed postmortem examinations.

#### REFERENCES

- (1) RABELLO, JR. Données nouvelles pour l'interprétation de l'affection de Besnier-Boeck: rôle de la lèpre. *Ann. Dermatol. et Syphilig.* **7** (1936) 571-597. (Reprinted in this number of *THE JOURNAL*, page 485)
- (2) MURDOCK, J. R. AND HUTTER, H. J. Leprosy: a roentgenological study. *American Jour. Roentgenol. and Rad. Therap.* **28** (1932) 598; reprinted in *Internat. Jour Lep.* **3** (1935) 339-344.