Boeck's Sarcoid*

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I do not believe there is a great deal to be gained by a review of the history of Boeck's sarcoid. I merely wish to remind you that it is being reported with increasing frequency and so must be considered in any chest diagnosis.

I wish to very briefly summarize some of the salient features regarding Boeck's sarcoid and then review a case that has been under my observation for several years.

ETIOLOGY

The various theories regarding the etiology only serve to demonstrate how much there is yet to be proved about this disease. Boeck originally considered the disease to be a constitutional one, caused by a non-virulent form of tubercle bacillus. This opinion is still held by a large number of men. A comparatively high proportion of cases will later develop an active tuberculosis. Pinner believes that tuberculous lesions develop as a part of the sarcoid lesions and not as a separate disease concurrent with the sarcoidosis. Kyrlé, Wende, and Goeckerman have demonstrated tubercle bacilli in early cases, and believe that if early lesions were examined more frequently the percentage in which tubercle bacilli are found would increase.

Others feel that it may be a nonspecific tissue response to various types of organisms, and perhaps to several organisms at the same time. In this connection, tubercle bacillus, leprosy bacillus, spirochaeta pallida, and Leishmania have been mentioned.

Pullünger and Ross and others have advanced a hypothesis that it may be a disease of the reticulo-endothelial system, comparable to Hodgkin's disease.

Kissmeyer and Nielsen believe that it is a chronic infectious granuloma. They advanced the theory that it may be due to a virus infection and that it is a specific pathologic entity.

PATHOLOGY

Histologically, the lesions resemble miliary tubercles. They are composed of epithelioid cells arranged in the form of miliary

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tubercles. These collections of cells may attain large proportions. They are not as a rule outlined by an inflammatory area of lymphoid cells. There is frequently a giant cell of the Langhans' type present in the center. Histologically, they closely resemble the lesion that Sabin has been able to produce in guinea pigs by injection of the phosphatide fraction of the tubercle bacillus. Rubin and Pinner report caseation in sarcoid lesions. In all the cases in which caseation was present, tubercle bacilli could be demonstrated in the lesion but not in the non-caseous lesion. The presence of a caseating lesion does not rule out the diagnosis of sarcoid.

BACTERIOLOGY

Pinner, in 1938, summarized the positive bacteriological findings that have been reported in the literature. There were very few. They did not prove conclusively that sarcoid was due to any one specific organism. In 1944, Pinner makes a statement that "failure to find tubercle bacillus in the majority of sarcoid lesions is not a convincing argument against their tuberculous etiology, and the presence of tubercle bacillus in some lesions is not a positive proof."

TUBERCULIN REACTION

In a great majority of cases there is a hyposensitivity to tuberculin or a complete anergy. In most cases after the development of clinical pulmonary tuberculosis, the tuberculin reaction becomes positive. Schaumann in 1936 reported that a negative skin reaction would become positive after the healing of cutaneous lesions in sarcoid. The theory has been advanced that the lesion produced an anticutin that neutralizes the tuberculo-pyrene. Pinner, in 1939, was unable to consistently demonstrate an anticutin that would account for the negative skin reaction.

CLINICAL FEATURES

This disease usually begins insidiously in early adult life, involving the skin, lymph nodes, bones, and lungs. It is extremely chronic. The clinical manifestations will depend on the structure that is involved.

Cutaneous Lesions—These usually consist of local areas of infiltration involving the skin of the cheeks, forehead, ears, arms, legs, fingers, toes, or back. They are usually of a dark red or brown color. The color is due to pigment deposit and not to increased vascularity. Pressure near the periphery of the lesion produces some blanching, but pressure in the center does not. Fusiform swellings of the fingers and toes are frequent, and there is usually an associated bone involvement.

Lymphadenopathy—In a large percentage of cases there is a
lymph gland involvement, which may be generalized or may be
confined to one or two sets of glands. The involved glands are
usually smooth, discrete and painless. Splenomegaly is frequent.

Pulmonary Lesions—Superficial, mediastinal, and peribronchial
nodes are usually enlarged. Frequently the pulmonary involvement
extends toward the bases, rather than towards the apices. Radiographically, the lungs show bilateral enlarged hilar glands. Atelectasis may be present as a result of compression by the hilar glands. There is usually interstitial fibrosis and multiple miliary foci resembling miliary tuberculosis.

Skeletal System—Small cysts are frequently found in the medullary portion of the bone. This involvement is most frequent in the bones of the fingers and toes. There is usually an area of decreased density in the bone with a surrounding area of increased density. These areas may be multiple and they may be very small.

Eye Lesions—In about ten per cent of the cases, involvement of the iris and ciliary body is present.

Parotid Glands—Enlargement of the parotid as the result of the presence of sarcoid has been reported. If iridocyclitis is present at the same time, a syndrome resembling uveoparotid fever is present.

Other Involvements—Lesions involving other organs as the kidney and spleen have been reported. In the kidney, the involvement is comparatively rare. In the spleen, it is quite frequently reported.

CASE HISTORY

I now wish to review a case that has been under my observation for several years.

This man was admitted to the sanatorium in December, 1942. At that time he was 33 years of age. He is white, has been married for ten years, and has two children that are living and well. His mother died of tuberculosis in 1916. Otherwise his family history is essentially negative. His past history is essentially negative.

This patient had a nocturia of several times a night for several years but did not consult a physician. There was no pyuria, dysuria, or hematuria. He was apparently in good health until the fall of 1941. There was a marked loss of weight that he could not account for at that time. During the winter he developed a dry hacking cough. In the spring he noticed that there was a definite loss of appetite. His cough, loss of weight and appetite persisted until the middle of the summer. A mild dyspnea developed. He lost 27 pounds. A tuberculin test at that time produced a negative reaction. He continued to work for approximately another month. At that time he had an attack of vomiting that was followed by nausea for several days. He had no previous gastro-intestinal symptoms that could have accounted for the vomiting. It did not seem to be associated with food and was not relieved by the usual alkali. He was advised to have his teeth examined and had one tooth extracted. Later there was another attack of vomiting and more teeth were extracted. The weight had dropped at this time from 217 pounds to 173. The sputum had gradually become productive of approximately an ounce of white,
somewhat thick sputum, in 24 hours. Dyspnea increased. In spite of the weight loss and lack of appetite, this patient continued to work. During an attack of coughing, he coughed up a plug of fibrin which was sent to a laboratory for examination. The report indicated a chronic lung disease and an x-ray was taken. This showed involvement in both lung fields. In spite of the negative tuberculin reaction, he was advised to have sanatorium treatment. About a month before his admission to the sanatorium, several small discrete movable non-tender glands appeared in the inguinal regions.

PHYSICAL EXAMINATION

Chest—Right: Essentially negative except for increased vocal fremitus anteriorly and posteriorly. There was some impairment of percussion note and no rales. Left: Essentially negative.

Glandular System—Negative except for freely movable discrete non-tender glands in both inguinal regions.


LABORATORY FINDINGS

Sputum—Approximately 25 cc. of serous sputum in 24 hours, negative for tubercle bacilli and fungus. Guinea pig inoculation negative for acid-fast organisms.

Stomach Washings—Negative for acid fast.

Blood—Sedimentation rate, 32 mm. in ½ hour; 76 mm. in 1 hour; N.P.N., 71.6; Hgb, 13½ grams; Rbc, 4,160,000; Wbc, 6,500; Poly, 60; Lymph, 25; Mono, 5; Eosin, 10.

Urine—A. M. Sample: Pus cells, 8-10; pus casts, 1-3; coarse gran. casts, 0-1; occasional fine gran. casts; rare hyaline casts; Mosenthal, 1005-1010; day, 999 cc.; night, 950 cc.; direct smear, negative for acid fast; culture, negative for acid fast.

Mantoux—0.01 mgm. (P.P.D.) negative. Patch test negative.

X-RAY FINDINGS

X-rays of the lungs showed large hilar glands on both sides with a productive type of infiltration in the first, second, and third interspaces anteriorly, on the right. There was a productive lesion in the first interspace anteriorly and in the third interspace on the left.

Repeated x-rays during this patient's stay in the sanatorium showed no essential change. X-rays of the hands were taken and one small cystic area of approximately 3 mm. in diameter was demonstrated.

Inguinal Glands were removed on both sides during patient's stay in the sanatorium. These were sent to different laboratories for examination. The report on all of these sections was consistent with a diagnosis of Boeck's sarcoi.

This man has been intensely interested in his case and in December, 1943, wrote me a rather detailed report of his condition since leaving the sanatorium. At that time he was working and apparently feeling quite well. Last April he had a rather severe cold, was in bed for eight days and lost 10 pounds in weight, but apparently recovered completely. There was still some dyspnea. X-rays of the lungs showed essentially no change. He still continues to have the kidney involvement. Urine showed approximately 1+ albumen. Blood pressure was 170/110. The inguinal glands are
still enlarged. A cervical gland became enlarged during the summer and this gland persisted. He has had several attacks of vomiting for no apparent reason, and in the interval between these attacks has had no trouble as far as the gastro-intestinal system is concerned.

After the first of the year (1944) he seemed to be in fair condition. Most of the joint aches and pains, as well as those of the left kidney region, disappeared. He was taking life fairly easy most of the time.

In March, 1944, he had a cold similar to the one in April, 1943, but not so severe. Chest felt congested. Much coughing and sputum. No fever or soreness of throat. General aches and pains, also kidney pains, returned and are still present. He has kept his appetite, however, and now is feeling quite well again. He has to be careful to prevent becoming tired. Inguinal glands have remained swollen and he is still bothered with coughing and shortness of breath.

He recently visited his doctor who found no change in the blood pressure.

He is now acting as an agent for an insurance company in his home county. He finds that he can do what he wants to do, just about when he wants to do it and keeps from becoming tired.

As I have stated, this man is still comparatively well. We have not had the advantage of having a pathological examination to find out definitely what type of involvement he has in the kidneys. It is my impression that this kidney condition very probably is sarcoid in nature, especially in view of the fact that he has had no previous history that would indicate any kidney involvement. I believe the attacks of vomiting have been on the basis of lack of kidney function, or it is possible that there could be a sarcoid involvement of the intestinal tract itself.

During his stay at the sanatorium, patient was treated symptomatically, and although there was some gain in weight, neither his x-rays nor his clinical condition changed appreciably. He was advised that the outlook was rather uncertain and that there was no treatment except for specific symptoms as they developed.

**SUMMARY**

A summary of the literature on Boeck's Sarcoid is presented. No definite conclusions can be drawn regarding the etiology or bacteriology. The pathology closely resembles that of tuberculosis. Clinically this disease is manifested by involvement of almost any part of the body. The usual involvement is in the glands, lungs, skin and bones. This disease is not incompatible with longevity. A case of Boeck's Sarcoid is presented in which there is probably kidney involvement. This patient is alive and able to carry on moderate work two years after diagnosis, and three years following the original symptoms.

**RESUMEN**

Se presenta un resumen de la literatura sobre la sarcoidosis. No se puede sacar conclusiones bien definidas acerca de la etio-
logía o la bacteriología. La patología es muy semejante a la de la tuberculosis. Desde el punto de vista clínico la enfermedad se manifiesta con la invasión de casi cualquiera parte del cuerpo. Comúnmente invade los ganglios, pulmones, piel y huesos. La enfermedad no es incompatible con la longevidad. Se presenta un caso de sarcoidosis en el que probablemente existe invasión del riñón. El paciente vive y trabaja moderadamente dos años después del diagnóstico y tres años después de la iniciación de los síntomas.

REFERENCES

   Kissmeyer and Nielsen, J. (1934).