Fatal Myocardial Sarcoidosis

A Case of Sudden Death

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The case history of a 47 year old white man who died suddenly is given. At autopsy extensive replacement of the myocardium by sarcoid granulation tissue was found. The lesions of sarcoidosis were also present in the lungs, the spleen, the hilar and mesenteric lymph nodes.

In recent years attention has been directed to the multiplicity of organs which may be affected in sarcoidosis. Originally described as a disease of skin and lymph nodes, sarcoid is now known to be capable of producing lesions in almost all the organs of the body. In 1929 sarcoid lesions in the epicardium were described by Bernstein and his colleagues. In 1937 Nickerson observed a case of fulminating sarcoidosis in which involvement of the myocardium and endocardium was found at autopsy. Since then a number of other cases have been recorded and death has been attributed to the myocardial lesion in at least eight cases; all of these occurred in Negroes, save one recorded by Yesner and Silver. This report dealt with the death of a 54 year old white male which was attributed to sarcoidosis of the myocardium. The present communication records the sudden death of a white man, aged 47, as a result of extensive involvement and replacement of the myocardium by sarcoid.

Case Report

R. C., a 47 year old white male, was admitted to Regina General Hospital on Dec. 21, 1950, and discharged on Jan. 30, 1951. He complained of admission of chest pain and shortness of breath for the preceding 24 hours. He stated that on the previous day he was stricken quite suddenly with a constricting pain beneath the sternum. The pain did not radiate but maintained its severity for three hours, after which it subsided somewhat; during this time the patient sat upright in a chair. He received a hypodermic injection 18 hours after the onset of pain and this provided some relief. He stated that he suffered a similar attack in the spring of 1950 while he was driving a tractor. The pain was so severe as to cause him to fall from the vehicle.

On admission he was a mildly obese man of good stature, complaining of slight nausea. The heart rate was 160 and the beats of irregular strength; gallop rhythm was present; blood pressure was 110/80. The heart did not appear enlarged on physical examination and no murmurs were heard. The radial pulse was 80 with a tendency to alternate strong and weak beats. The lungs appeared clear. The remainder of the physical examination was unremarkable. A chest film on December 27 revealed no bony abnormalities of the thorax. The right diaphragm was considerably elevated and there were some irregular minimal streaky shadows across the right lung base. The heart shadow appeared normal. On January 26 another chest plate was made and the right diaphragm had come down nearly to normal level. The infiltration previously seen in the right base had disappeared. Some very faint infiltration was seen, however, in the outer right second and third intercostal spaces which was not present on the previous film. The cardiovascular shadow appeared to be within normal limits. The impression at that time was of a right basal lesion which had resolved with the presence now of a new lesion in the upper third of the right lung field. An electrocardiogram (fig. 1) was taken and showed evidence suggestive of myocardial and subendocardial ischemia.

The clinical impression was one of coronary thrombosis and the patient was treated with anti-coagulants which were begun on December 22 and discontinued on January 15.

The hospital course was uneventful. The patient was confined to bed until January 3 at which time he was allowed bathroom privileges. He had no recurrence of his chest pain and appeared to be making satisfactory progress. On January 21 he was allowed to sit up for a short time and this privilege was gradually extended until his discharge from the hospital. At the time of his discharge he appeared to be considerably improved.

On February 9 he had some teeth extracted under anesthesia and appeared to tolerate this procedure well. On February 13 he was found dead in a hotel.

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room. He had evidently died suddenly as he had been seen in apparently good health shortly before his death. His death was the subject of a coroner's investigation.

Autopsy Findings (ML-5-51): The autopsy was performed 17 hours after death. The body was that of a well nourished, middle-aged man of good nutrition. The skin surface appeared normal.

The lungs were voluminous and heavier than normal; right weighed 850 Gm., left 780 Gm. Numerous adhesions were present between the visceral and parietal pleural surfaces. On palpation the lung texture felt coarse. An extensive dissection of the bronchial system was undertaken but no evidence of primary neoplasm was found. The lungs were not conspicuously pigmented, and there was no evidence of pneumonia. Moderate pulmonary edema was present.

The heart weighed 430 Gm. The pericardial sac appeared healthy and contained 20 ml. of clear yellow fluid. Inspection of the heart before dissection revealed numerous white spots on the anterior and posterior surfaces of both ventricles and auricles. These did not have the appearance of milk spots and on section proved to be fibrous tissue. The fibrous tissue, however, appeared somewhat more granular than normal, and its distribution was quite irregular; it bore no relationship to the coronary artery system. Similar lesions were noted in the papillary muscles. The lesions encroached on the endocardial surface of the heart. The largest fibrous area was present fairly high up on the posterior wall of the left ventricle under the posterior cusp of the mitral valve, and much of the anterior wall of both right and left ventricles was replaced. The endocardium and valves themselves were unremarkable. The coronary arteries were comparatively free from atheroma. The aorta showed moderate atheroma together with some fatty streaking of the intima.

The peritoneal cavity appeared healthy. The gastrointestinal tract, liver, gallbladder and biliary passages, pancreas, adrenals, kidneys and testes all appeared healthy.

The spleen was considerably enlarged, weighing 590 Gm. Section revealed the bulk of the parenchyma to be normal in color and soft in texture. Scattered throughout, however, were several small white, firm, circumscribed nodules, the largest measuring 1.5 cm. in diameter. The texture of these nodules was indistinguishable from that of the fibrous tissue in the myocardium.

Enlargement was noted of the hilar lymph nodes and of the mesenteric lymph nodes. The texture of the nodes was soft.

The skull and brain were healthy. The pituitary was normal. X-rays of the long bones and the digits, taken at autopsy, revealed no abnormality.

Cause of Death: At autopsy the cause of death was thought to be acute heart failure resulting from extensive fibrous replacement of the myocardium.

Histologic examination of the lesions in the myocardium revealed them to be areas of fibrosis in which multinucleate giant cells were abundant (figs. 2, 3 and 4). The connective tissue was sparsely infiltrated by lymphocytes but, on the whole, avascular and collagogenous, and the giant cells lay scattered irregularly in it. In a few areas the giant cells lay in clumps. Some of these cells contained rounded Schaumann bodies but no asteroid inclusions were seen. In a few areas the connective tissue
Fig. 3. The edge of the lesion showing more detail. (X 120.)

Fig. 4. A cluster of multinucleate giant cells. (X 120.)

Fig. 5. A sarcoid follicle in a lymphatic in the lung. (X 120.)

Fig. 6. A sarcoid follicle in the spleen. (X 100.)
was more cellular and showed a tendency to whirling and the formation of tubercle-like nodules, but in the myocardium these were not numerous.

The lung showed an interesting picture (fig. 5). Follicular lesions composed of endothelioid cells with numerous giant cells were present in the peribronchial and perivascular lymphatics. No gross areas of fibrosis were seen.

The spleen was congested and cellular. The sinus lining cells were more prominent than usual. The pale nodules were fibrous areas similar to those in the myocardium. In this region, however, the connective tissue was more cellular and follicles were more numerous. Giant cells were abundant (Fig. 6).

The hilar and mesenteric lymph nodes were extensively replaced by a granuloma of follicular architecture. The follicles were composed of endothelioid cells and numerous giant cells.

Histologic examination of the liver, kidney, pancreas, adrenals, thyroid and pituitary failed to reveal the presence of the characteristic follicles. Stains for the tubercle bacillus were negative. Periodic acid Schiff stains failed to reveal any evidence of fungus. In none of the lesions was there any caseation.

The histologic picture was compatible with a diagnosis of sarcoid. It is generally agreed that the diagnosis of this disease can be made only histologically and must then fulfill certain criteria.11

SUMMARY

A case is presented of a 47 year old white man who died suddenly. At autopsy, extensive replacement of the myocardium by granular fibrous tissue was found. Histologic examination of the myocardium revealed the presence of sarcoidosis. Sarcoid lesions were also present in the spleen, in the perivascular and peribronchial lymphatics of the lung and in the mesenteric and hilar lymph nodes.

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SUMARIO ESPAÑOL

Se informa el caso de un paciente blanco de 47 años de edad que murió subitamente. En la autopsia se encontró un reemplazo extensivo del miocardio por tejido fibroso granular. Examen histológico del miocardio reveló la presencia de sarcoidosis. Lesiones sarcoides también se encontraron en el vaso, en los linfáticos perivasculares y peribronquiales del pulmón y en los ganglios mesentéricos y del hilo.

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