Sudden death associated with compression of pulmonary arteries in sarcoidosis

Justin M.E. Martin, MB, BCh, FRCPC
Graeme P. Dowling, MD

Pulmonary and hilar lymph node involvement occurs in 80% to 90% of cases of sarcoidosis. In most instances pulmonary sarcoid granulomas are perivascular; they may involve the walls of small and medium-sized vessels to produce granulomatous vasculitis.[1-3] To date there have been only six reports of external compression of major pulmonary arteries by enlarged hilar lymph nodes in sarcoidosis;[1-6] none of these cases was associated with sudden death. We report the first known case of sarcoidosis wherein such compression was thought to be a major contributing factor in the sudden death of a previously healthy young man.

Case report

A 27-year-old male jogger was found dead on a sidewalk early in the morning. He had jogged 6 km regularly three to four times per week. He was reported to have been healthy until his death, his only recent complaint having been of excess tiredness after jogging. He had had no complaints of chest pain, dyspnea or any other discomfort and had only seen his family physician for minor ailments.

Significant autopsy findings were confined to the chest. Bilaterally the hilar lymph nodes were significantly enlarged, up to 3 cm in diameter, and this had resulted in significant external compression of the left and right main pulmonary arteries; the lumen of the left pulmonary artery was reduced to a slit (Figs. 1 and 2). Sections of the nodes had a homogeneous tan appearance; some areas showed a fine nodular configuration. A subpleural

From the departments of Pathology, Foothills Hospital, Calgary, and University of Calgary

Reprint requests to: Dr. Justin M.E. Martin, Department of Pathology, Foothills Hospital, 1403-29th St. NW, Calgary, Alta. T2N 2T9

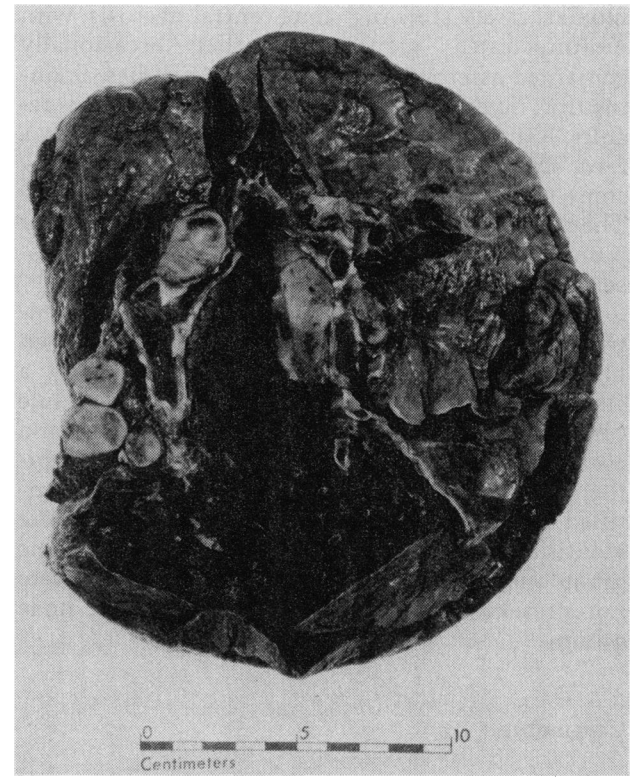

Fig. 1—Prominent hilar lymphadenopathy in left lung.

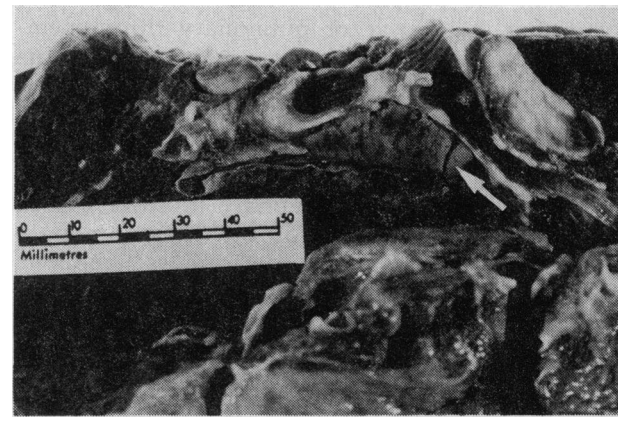

Fig. 2—Hilum of left lung. Note that markedly enlarged lymph node (arrow) has reduced lumen of pulmonary artery to slit.

focus of fine granular consolidation, 2 cm in diameter, surrounded by several smaller firm, brown nodules, was present in the lower lobe of the left lung. The heart weighed 315 g. The right ventricular wall was 0.3 cm thick and the left 1.3 cm at its widest point. The right coronary artery was normal, but the left anterior descending coronary artery contained an atheromatous plaque that occluded 50% of the lumen. Careful external examination and serial sectioning of the ventricular conducting system revealed only a pale subendocardial nodule, 1.5 mm in diameter, in the septum, adjacent to the right ventricle.

Histologically, the hilar lymph nodes and the pulmonary nodules showed the features of sarcoidosis: numerous noncaseating epithelioid granulomas, rarely showing some central necrosis, with multinucleated giant cells that occasionally contained asteroid and Schaumann's bodies. Staining for mycobacteria and fungi had negative results. Random sections of the lungs showed only a few scattered microscopic sarcoid granulomas, some of which involved small pulmonary vessels. There was no evidence of pulmonary fibrosis or pulmonary hypertension: the intrapulmonary vessels were of normal dimensions.

Histologic examination of the entire interventricular myocardium, which had been removed en bloc, revealed only minimal involvement by a similar process. The pale subendocardial nodule consisted of a mixed inflammatory response and associated giant cells. A second noncaseating epithelioid granuloma, also subendocardial, was identified in a random section. Rare granulomas were identified within the pericardial fat. Microscopic granulomas were also found in the liver but were not present in the spleen or other lymph node groups.

Comments

Symptomatic compression of the major pulmonary arteries by hilar lymph nodes in sarcoidosis is rare, having been reported in only six cases. Three of the patients presented with signs and symptoms suggestive of pulmonary thromboembolism (pulmonary angiograms delineated the true cause),[1,2,4] one presented with pulmonary hypertension thought to be due to fibrous mediastinitis induced by sarcoidosis,[5] one presented with weakness due to sarcoid myopathy and was found to have a harsh systolic pulmonary murmur caused by external compression of the left pulmonary artery by enlarged hilar lymph nodes,[6] and one died of chronic cor pulmonale with widespread pulmonary sarcoidosis and compression of the left pulmonary artery by hyalinized lymph nodes.[4]

Our report seems to be the first of sudden death associated with pulmonary artery compression by hilar lymph nodes in sarcoidosis. Despite the compression of both main pulmonary arteries there was no microscopic evidence of pulmonary hypertension. However, we think that the combination of strenuous exercise, significant but localized coronary atherosclerosis and pulmonary arterial compression induced a fatal arrhythmia.

Sudden death is an uncommon initial manifestation of sarcoidosis[7] and in most cases has been associated with extensive sarcoid involvement of the myocardial conducting system. In our case only two small subendocardial foci of involvement by sarcoid were found. The nodal conducting system was not examined histologically, so we are unable to rule out involvement there, but it is unlikely in view of the minor degree of demonstrated myocardial involvement.

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