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Case report

Sudden death due to cardiac sarcoidosis in a case of suspected homicide

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Summary. In a case of suspected homicide death due to natural causes – cardiac death (SCD) – was found at autopsy. Despite an extensive replacement of myocardial tissue by sarcoid granulomata there was no history of cardiac dysfunction or preceding symptoms. The transmurally infiltrating granulomata and the concomitant fibrosis were predominantly confluent. They occupied vast areas within the interventricular septum and the adjacent posterior wall of the left ventricle. The only other organs involved were mediastinal lymph nodes, which appeared macroscopically normal.

Key words: Cardiac sarcoidosis – Sudden death – Sarcoid heart disease

Zusammenfassung. Es wird über einen plötzlichen Herztodesfall berichtet, bei dem zunächst ein Fremdverschulden nicht ausgeschlossen werden konnte. Die Sektion ergab eine Sarkoidose des Herzens mit einer umfangreichen teils transmuralen granulomatösen Durchsetzung des Myokards in der Hinterwand des linken Ventrikels sowie im Kammerseptum. Abgesehen von einigen mediastinalen Lymphknoten gab es keinen weiteren Organbefall.

Schlüsselwörter: Plötzlicher Herztod – Sarkoidose – Sarkoidose des Herzens – Plötzlicher Tod

Case history

A 34-year-old male was found dead on the premises of a building construction site. Two young unemployed males, who admitted having discovered and robbed the allegedly lifeless body were held as suspects for homicide. Because of the disarray of the clothing and the surroundings, as well as the extreme congestion and cyanosis of the face, violence could not be excluded. Later investigations confirmed that the deceased had never been seriously ill and had no previous history of cardiac dysfunction.

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Major autopsy findings

The sun-tanned athletic body showed no external injuries or other changes suggestive of the application of force. The face was markedly congested and cyanotic. The jugular veins were prominent on both sides of the neck. There were no external signs suggestive of strangulation. Acute passive congestion was present in the lungs, the liver, the spleen and the leptomeningeal blood vessels as well as within the cerebral medulla. The liver showed moderate fatty changes with a nutmeg appearance of the cut surface. The lymph nodes were of normal size. The moderately enlarged heart weighed 450 g and showed a concentric hypertrophy of the left ventricle. The upper half of the left ventricle immediately below the aortic valve showed a well demarcated greyish subendocardial thickening reminiscent of fibrosis which extended into the ventricular septum (Fig. 1a). A similar subendocardial change was observed at the corresponding opposite side of the interventricular septum – viewed from the right ventricle (Fig. 1b). Parallel axial sections showed transmural and transseptal replacement of myocardial tissue by apparently nodular lesions which were partly confluent (Figs. 2a and 2b).

Histology

Histological examination revealed extensive replacement of myocardium by numerous partly confluent granulomata without central necrosis or caseation. Epitheloid cells and multinucleated giant cells were abundant (Fig. 3). Laminated basophilic inclusion bodies – Schaumann bodies – were occasionally present. Areas of involuted granulomata showed extensive and dense fibrosis.

Similar granulomata were observed in the lymphatic nodes from the mediastinum which otherwise appeared innocuous.

Fluorescence microscopy of standard 5 μ sections stained with acridine orange and examined under ultraviolet light showed areas of various shades of greenish

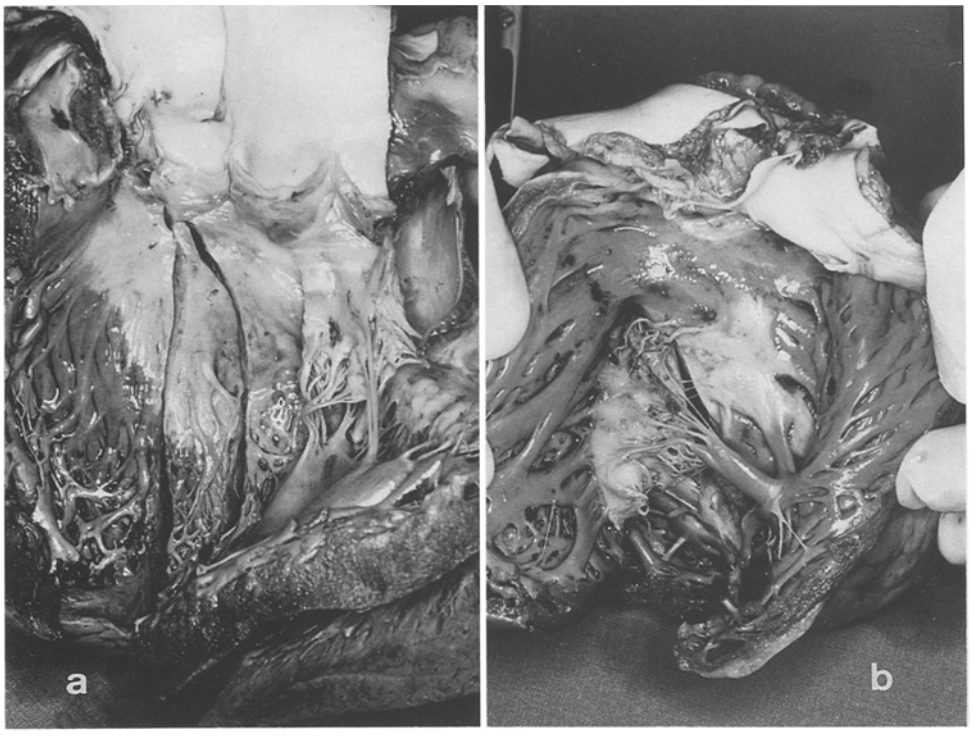

Fig. 1a, b. Left ventricle (**a**) well demarcated greyish subendocardial thickening involving the upper halves of the posterior left ventricular wall and the interventricular septum. Right ventricle (**b**) corresponding right ventricular side of the interventricular septum showing a similar well demarcated subendocardial thickening

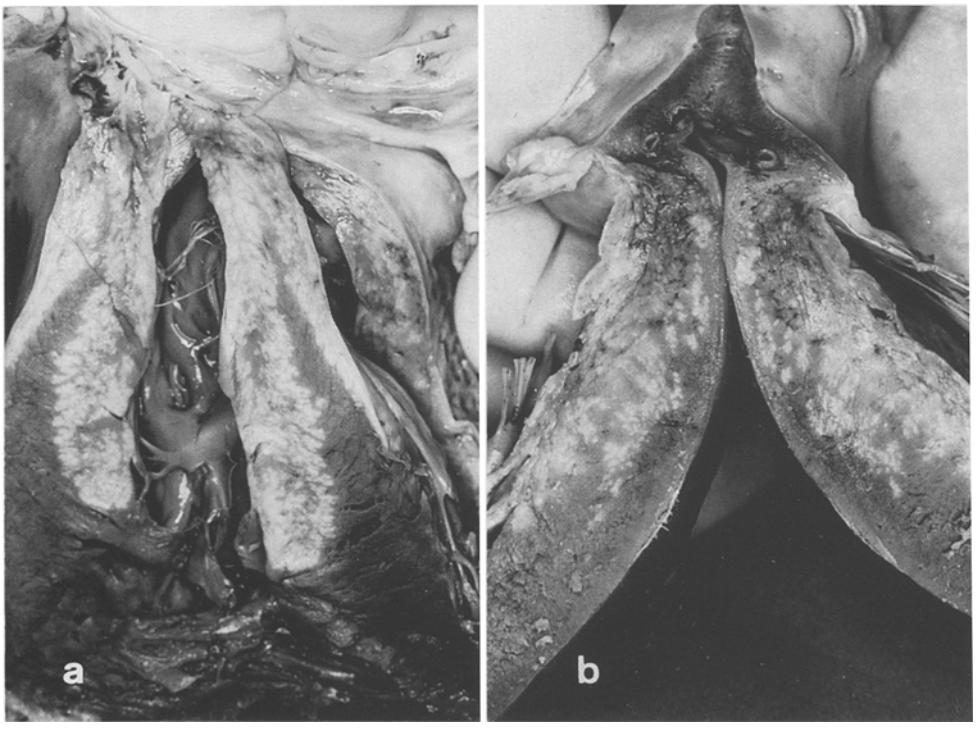

Fig. 2a, b. Axial sections showing transmural involvement of septum (**a**) and posterior wall of left ventricle (**b**) by predominantly confluent granulomata with concomitant fibrosis

fluorescence in the immediate vicinity of several granulomata [1–3]. The myocardium in areas distant from the granulomata showed a gold-brown fluorescence.

Immunhistochemistry

Monoclonal antibodies against T- and B-lymphocytes were used to identify and differentiate the lymphocyte population around the granulomata. They proved to be mainly T-lymphocytes and only occasionally B-lymphocytes.

Discussion

Sudden and unexpected death due to natural causes is an important diagnosis in forensic case work especially

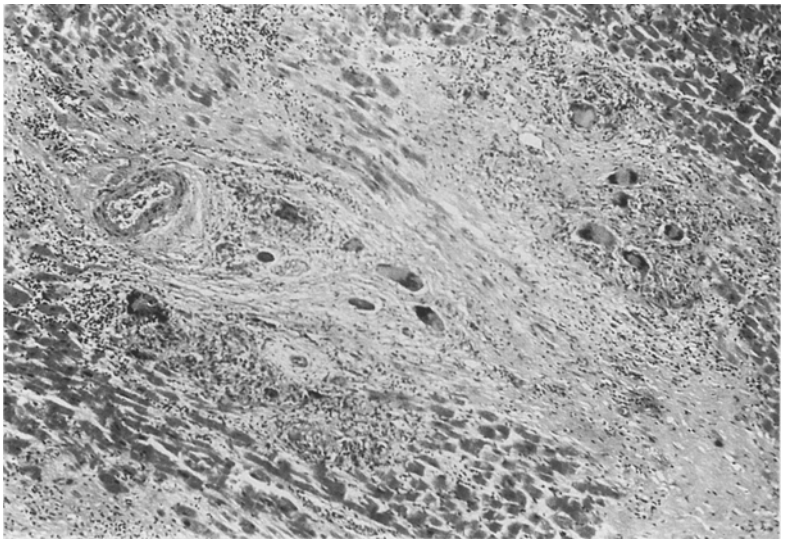

Fig. 3. Extensive replacement of myocardial tissue by confluent granulomata and fibrosis. No central necrosis, no caseation. Abundant epitheloid cells and multinucleated giant cells. (H & E.; *magnification* 180 ×)

when the deceased has otherwise been healthy. In the majority of such cases cardiac involvement has been a frequent finding at autopsy, whereby ischaemic changes appear to be particularly prominent. Next on the list in our experience has been myocarditis. Cardiac sarcoidosis, although subject of a number of clinical and clinicopathological reports, is rarely mentioned in forensic medical case reports in connection with sudden unexpected death. This could be because of the more frequent extra cardiac manifestation and the observation that there seems to be a negative correlation between cardiac involvement and the involvement of other organs [4].

Depending on the amount of the replaced myocardial tissue, the extent of scarring and the location of the granulomata as well as the degree of fibrous replacement of involuting granulomata, various forms of cardiac dysfunction can occur [5–9]. These include atrioventricular block, bundle branch block, ventricular arrhythmia, congestive heart failure etc. The arrythmogenic tendency of cardiac sarcoidosis and the electrocardiographic evidence of myocardial damage has sometimes led to the initial diagnosis of myocardial infarction. In the vast majority of the reported cases, usually with preceding symptoms, the granulomata were found mainly in the left ventricular posterior wall and in the ventricular septum [5, 10, 11, 12]. Sarcoid infiltration of the sinus node and the A-V node has also been reported [13, 14].

Although devoid of symptoms, the granulomatous changes found in this case were similarly distributed (Fig. 1). The topography and the intramural distribution of these obviously chronic changes within the ventricular septum clearly suggested an involvement of the conduction tissue system. Unlike most of the reported cases, the changes in this case apparently did not cause any conduction disturbances or cardiac dysfunction of an equally chronic nature in spite of the long-standing extensive replacement of myocardial and possibly of conduction tissue. This observation is further supported by the abscence of gross pathological and histological evidence of chronic congestion in the lungs and other organs.

In a number of cases of sarcoid heart disease the first discovery of the disease has been at autopsy [5, 14, 15, 16] because the individuals showed no symptoms whatsoever. Only in a few such completely asymptomatic cases have there been reports on the extent, topography and severity of the involvement of the myocardium and the conduction system [14]. In these cases (apparently healthy individuals, who had lived for several years with these pathological changes confined to the heart), recent additional precipitating factors most likely play a decisive role and are more or less assumed. The shifts in the secondary fluorescence pattern of the myocardium, as found in some of the perifocal areas in this case, suggest some form of change (possibly recent) since tissue areas distant from the granulomata uniformly retain the gold brownish fluorescence of normal myocardium [1–3]. It has been pointed out that even in non-SCD cases agonal ischaemia may be sufficiently protracted to produce such shifts as an indication of recent myocardial injury [17]. Such demonstrable changes could be seen as the final precipitating factors which are superimposed upon the preceding clinically inapparent but morphologically severe sarcoid infiltration.

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