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CASE REPORT

Sarcoidosis mimicking ischaemic ventricular arrhythmia and pulmonary embolism

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ABSTRACT

Sarcoidosis is a multisystem granulomatous disorder characterised pathologically by the presence of noncaseating granulomas in the organs involved. Cardiac involvement, although well known, is rare.
We describe a 72-year-old patient who was admitted to the intensive care unit after coronary artery bypass grafting. She developed refractory right and left ventricular failure complicated by multiple organ failure and died three days later. Postmortem examination revealed extensive sarcoidosis. On hindsight, preoperative ventricular tachycardia and an abnormal perfusion-ventilation scintigraphy of the lungs were manifestations of an underlying sarcoidosis.

INTRODUCTION

Sarcoidosis is a multisystem granulomatous disorder of unknown aetiology characterised pathologically by the presence of noncaseating granulomas in the organs involved.[1] Most frequently sarcoidosis involves the lung. Extrapulmonary sarcoidosis can affect all organ systems. Granulomatous involvement of the heart may lead to various cardiac problems especially arrhythmias. Nevertheless cardiac involvement only gives rise to clinical manifestations in 5% of the patients.[2,3] Initial cardiac presentation of sarcoidosis is rare.[4] We report a patient who underwent coronary artery bypass grafting (CABG) because of symptomatic single-vessel disease and recurrent ventricular tachyarrhythmia who was found to have extensive sarcoidosis with involvement of the heart.

CASE REPORT

A 72-year-old woman was admitted to our intensive care unit after coronary artery bypass grafting of the obtuse marginal branch. Four months before admission a diagnosis of pulmonary embolism was made after she had presented with dyspnoea and pleuritic pain. The diagnosis of pulmonary embolism was based on a positive D-dimer and a high probability mismatch on nuclear perfusion ventilation scanning. Her previous medical history was unremarkable apart from an asymptomatic left-sided carotid stenosis.
Three months before admission she presented with chest pain and ventricular tachycardia that converted to sinus rhythm after amiodarone therapy. Subsequent cardiological evaluation showed normal left and right ventricular function on echocardiography. ECG showed sinus rhythm with right bundle branch block.
Coronary angiography revealed an occluded obtuse marginal branch while the other coronary arteries were normal. Radionuclide imaging (myocardial scintigraphy, TC-99M MIBI) showed an irreversible posterolateral defect. There were no segmental areas of decreased uptake of the ventricular myocardium corresponding to areas of fibrogranulomatous replacement. Initially percutaneous coronary intervention was intended. This turned out to be technically impossible and subsequently the patient was scheduled for an off-pump CABG. During cutdown of the left internal thoracic artery (LITA), the patient developed a refractory cardiogenic shock, needing extracorporeal circulation. The LITA was very small with an almost absent flow and considered unsuitable. An aorto-coronary venous bypass graft was constructed to the marginal branch. The patient could be easily weaned from bypass without signs of ischaemia.

Postoperatively the patient was on mechanical ventilation with normal bilateral breathing sounds. Haemodynamically she was stable. Swan-Ganz pressure tracings recorded elevated pulmonary artery pressures and a low cardiac index (*table 1*). There were no enlarged lymph nodes and no pathological findings on abdominal examination. There was slight peripheral oedema at the extremities. Laboratory findings directly postoperatively were a haemaglobin of 6.5 mmol/l, thrombocyte count 125 x 10.9/l, leucocyte count 14.4 x 10.9/l, partial thromboplastin time (PTT) 1.82 and activated PTT 67. Electrolytes, liver and renal function tests were normal. Arterial blood gas analysis was unremarkable. Despite the low cardiac index the patient was extubated after initial postoperative care and on the first postoperative day she was discharged to the regular ward. In the next three days the patient complained of increasing dyspnoea and chest pain. Transthoracic echography showed pericardial effusion and a decreased right ventricular function. Both right atrium and ventricle were enlarged. A diagnosis of pericardial tamponade was made and rethoracotomy followed. During this procedure pericardial fluid was removed. Postoperatively the patient was re-admitted to the ICU.

In the postoperative phase the patient developed cardiac shock. Swan-Ganz measurements showed high pulmonary artery and central venous pressures and a low cardiac index (*table 1*). Despite therapy the patient developed multiple organ failure and died the following day.

The postmortem examination showed a hypertrophic heart with dilatation of the right ventricle (*figure 1*), and an old infarction and fibrosis of the left ventricle. The graft was open. Surprisingly, microscopic examination revealed myocardial sarcoidosis (*figure 2*), and extensive granulomas in the mediastinum, liver and lungs. Tuberculosis as well as fungal infection was excluded using polymerase chain reaction. There was no evidence of pulmonary embolism.

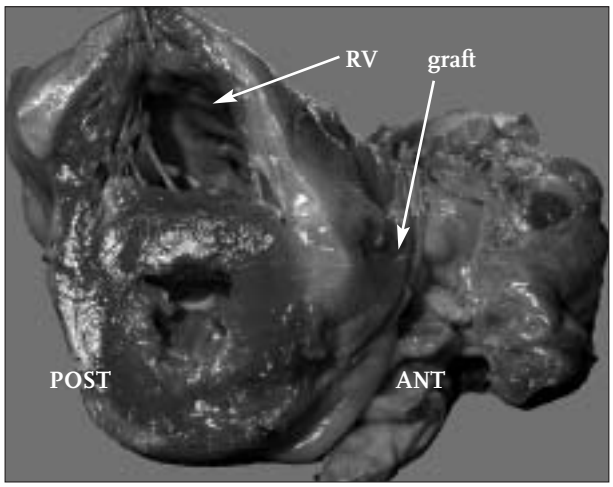

Figure 1
Cardiac hypertrophy (weight 430 g) and dilatation right ventricle
RV = right ventricle; post = posterior; ant = anterior.

DISCUSSION

Sarcoidosis is a systemic disorder of unknown cause which is characterised by noncaseating granulomas in the organs involved.[1] Although the lung is usually involved, the disease is known for its extrapulmonary manifestations.[5]

Table 1
Preoperative and postoperative haemodynamic parameters (after CABG and re-thoracotomy)

PARAMETERS (REFERENCE VALUE)	PREOPERATIVE	POST-CABG	POST-RETHORACOTOMY
Heart rate	90	63	112
Systolic BP (mmHg)	145	118	108
Diastolic BP	57	50	65
Mean BP	88	74	83
CVP (1-6 mmHg)	5	4	10
PAP systolic (15-28 mmHg)	40	41	57
PAP diastolic (5-15 mmHg)	20	17	30
PAOP (6-12 mmHg)	11	10	15
CI (2.4-4.0 l/min/m^2)	-	1.7	1.5
SVRI (1600-2400 dynes.sec. m^2/cm^5)	-	3388	3972
PVRI (200-400 dynes.sec. m^2/m^5)	-	774	1453
LVSWI (40-60 g.m/m^2)	-	26	15
RVSWI (4-8 g.m/m^2)	-	9	7
SV (60-70 ml/stroke)	-	41.3	13

BP = blood pressure; CVP = central venous pressure; PAOP = pulmonary artery occlusion pressure; PAP = pulmonary artery pressure; CI = cardiac index; SVRI = systemic vascular resistance index; LVSWI = left ventricular stroke work index; RVSWI = right ventricular stroke work index; SV = stroke volume.

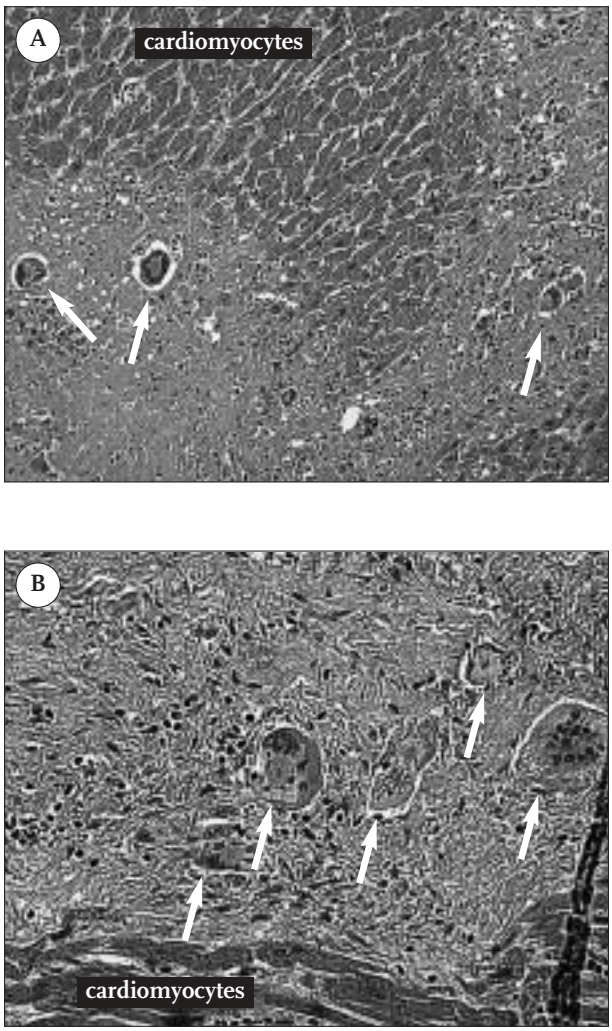

Figure 2
Myocardial sarcoidosis (A: lateral wall, B: posterior wall, cardiomyocytes and area with noncaseating granulomas, giant cells (arrow) and fibrosis

Cardiac manifestations of the disease are rare and reported in up to 5% of the patients with sarcoidosis.[2,3] It is unusual for sarcoidosis to present with isolated cardiac involvement.[4] Autopsy studies show a higher percentage of myocardial sarcoidosis. In an autopsy study, cardiac involvement proved to be the cause of death in 37% of the patients with sarcoidosis. In only 45% the diagnosis of sarcoidosis was suspected or established antemortem.[6] Cardiac sarcoidosis may cause interstitial inflammation which initially impairs diastolic function, whereas systolic function remains normal or nearly normal.[7] Subsequent inflammation and fibrosis result in impaired systolic function. Diffuse hypokinesia can occur, as well as focal abnormalities of regional wall motion that especially affect the basal septum but spare the apex.[8] The course of the disease is variable; in some patients it progresses rapidly to death with no preexisting symptoms.[9-11]

Half of the patients with cardiac sarcoidosis have electrocardiographic abnormalities of rhythm conduction and repolarisation. Clinical manifestations depend upon the location and extent of inflammation and should especially be suspected in young patients with known sarcoidosis presenting with arrhythmias. Involvement of the ventricular septum and conduction system may lead to a variety of arrhythmias and sudden death. Sudden death due to ventricular tachyarrhythmias or conduction block accounts for 25 to 65% of the deaths due to cardiac sarcoidosis.[12-14] Chronic pulmonary hypertension and cor pulmonale result from inflammation and subsequent severe scarring of the pulmonary parenchyma and vascular obliteration. In this setting death from sarcoidosis commonly results from right ventricular failure. The various clinical manifestations due to cardiac involvement are presented in *table 2*.

Table 2
Manifestations of cardiac sarcoidosis

Conduction abnormalities	First-degree heart block Intraventricular conduction defects Complete heart block
Ventricular arrhythmias	Abnormal automaticity Disrupted ventricular activation and recovery Sustained or nonsustained ventricular tachycardia
Supraventricular arrhythmias	Ectopic atrial activity Paroxysmal atrial tachycardia Atrial flutter/fibrillation
Heart	Systolic dysfunction Diastolic dysfunction Ventricular aneurysm
Valvular dysfunction	Mitral incompetence due to papillary muscle involvement
Simulated infarction	Transmural, non-Q-wave
Pericarditis	Rare, detected by echocardiography
Cor pulmonale, right-sided heart failure	Due to advanced pulmonary sarcoidosis

Sarcoidosis may mimic pulmonary embolism. Nuclear imaging (V/Q scan) can be falsely interpreted as pulmonary embolism.[15-18] Moreover D-dimer concentrations may be elevated in pulmonary sarcoidosis.[19-21]
Cardiac sarcoidosis may cause cardiomegaly and heart failure but may be difficult to establish. Firm diagnostic tests are not available and the diagnosis can only be established on the combined diagnostic modalities available and the exclusion of (other) structural heart disease. Although the prognosis of symptomatic cardiac sarcoidosis is not well defined, treatment with corticosteroids seems to delay the progression of inflammation and fibrosis.[1,12,14] Pacemakers are indicated when evidence of high-grade conduction disease is present. Automatic implantable cardioverter-defibrillators (AICD) are recommended in survivors of sudden death or patients with refractory ventricular tachyarrhythmias.[22-24]

CONCLUSION

Retrospectively, a high index of suspicion for a noncoronary explanation of the chest pain and the arrhythmia could have placed the ventricular tachycardia and the abnormal V/P scan in a different perspective: both fit the diagnosis of sarcoidosis, especially while there was single-vessel coronary stenosis. Even in retrospect there was little evidence of sarcoidosis in our patient preoperatively.
In conclusion, sarcoidosis is often not diagnosed nor suspected antemortem. The combination of ventricular arrhythmia, pulmonary hypertension, abnormal V/P scan and a positive D-dimer may be a clue to the right diagnosis

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