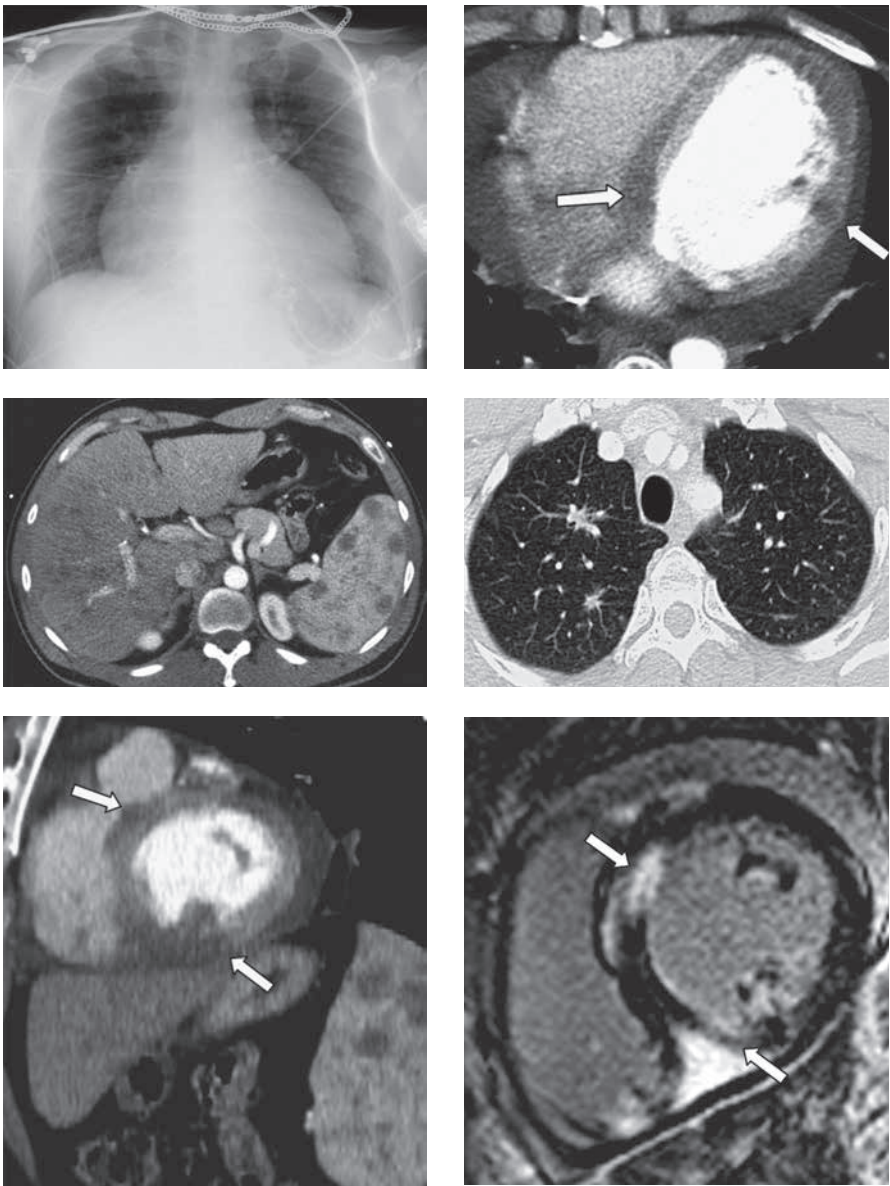


MULTISYSTEMIC SARCROIDOSIS WITH CARDIAC INVOLVEMENT

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Key-word: Sarcoidosis

Background: A 42-year-old male presented with dyspnea following a syncopal episode a few hours before admission. He had completed his military service 2 months previously. Electrocardiogram showed a complete heart branch block with severe bradycardia measured at 30 bpm. Imaging included chest radiography, CT scan of the chest, heart and abdomen and MRI of the heart.



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Fig.	3	4
	5	6

Work-up

On bedside frontal chest radiograph (Fig. 1), cardiac enlargement is noticed.

Axial non ECG-gated contrast-enhanced CT scan, section through the heart (Fig. 2) shows hypodense wall thickening near the cardiac apex and hypodense nodular lesions in the lateral wall of the left ventricle, and at the base of the interventricular septum (arrows). A pericardial effusion is also noted.

Contrast-enhanced abdominal CT scan (Fig. 3) demonstrates multiple irregularly shaped hypodense nodules of variable size in liver and spleen.

On chest CT scan (lung window setting) (Fig. 4), two areas of interstitial peri-bronchovascular thickening in the right upper lobe are seen.

On short-axis cardiac reformatted CT image (Fig. 5), hypodense lesions of the interventricular septum and the inferior wall of the left ventricle (arrows) are observed. Hypodense lesions are also seen in the spleen.

Contrast-enhanced turbo FLASH short-axis cardiac MRI (Fig. 6) demonstrates late enhancement (white stain) (arrows) in the same areas as that of the hypodense lesions in Fig. 5. Topography of the enhancement is variable: subendocardial, subepicardial, midmyocardial or transmural. Normal myocardium presents in dark stain.

Radiological diagnosis

Based on the radiological and the ECG findings, the diagnosis of *multisystemic sarcoidosis* including cardiac involvement was made. Sarcoidosis was subsequently confirmed by transbronchial lung biopsy.

Discussion

Sarcoidosis is a systemic disorder of unknown cause that is characterized by non caseating granulomas with proliferation of epithelioid cells. The most common radiologic findings are bilateral hilar lymphadenopathies, often associated with pulmonary infiltrates. However, extrathoracic involvement can be an initial manifestation including lesions of the skin, eyes, liver, spleen, parotid glands, central nervous system, genitourinary system, muscles and bones.

Cardiac involvement was observed in 25% of patients in an autopsy series, whereas clinical evidence of cardiac sarcoidosis (CS) has been reported in only 5% of all patients with sarcoidosis. Cardiac sarcoidosis is important to diagnose as it may present with heart branch block or congestive heart failure. It is estimated that, in Japan, nearly 80% of patients who die of sarcoidosis do so from cardiac involvement. Sudden cardiac death may be the presenting or an early feature of the disease. CS must be considered as a main differential diagnosis when a young patient presents with complete heart branch block. Other causes of heart branch block, such as Lyme disease, need to be excluded because these may be reversible with therapy and may not require permanent pacing.

Results from endomyocardial biopsy may frequently be negative because of the patchy distribution of the granulomas. Blood tests including hypercalcemia or elevated serum angiotensin converting enzyme (ACE) are non specific and often non diagnostic. Occasionally, echocardiography will reveal findings consistent with, but not pathognomonic for disease. Diagnosis of CS is further complicated by the absence of an evidence-based diagnostic approach so far. Thallium and Gallium-67 scintigraphies have been used in the past for the evaluation of patients with suspected CS, but nowadays they have become less frequently due to their lack of sensitivity and specificity. Most cases of CS are confirmed by using various imaging tests such as contrast-enhanced cardiac MRI and PET scan. Recent studies suggested that PET may be more sensitive than MRI, whereas MRI may have a higher specificity. Unlike PET, MRI does not expose patients to ionizing radiation. On MRI, the distinction between ischemic and non-ischemic myocardial damage can be made on the basis of pattern and distribution of late enhancement within the myocardium. In myocardial infarction scar, late enhancement always involves the subendocardial layer. If, on the other hand, late enhancement respects the subendocardial layer, different nonischemic myocardial diseases have to be considered. In our patient topography of the lesions was transmural or subendocardial, but also subepicardial and midmyocardial. Little has been written on the role of CT in the diagnosis of CS so far. In our patient, the severe bradycardia (30 bpm) permitted the visualisation of the myocardial lesions without the use of ECG-gating. The simultaneous findings of liver, spleen and pulmonary involvement helped to suggest the final diagnosis of multisystemic sarcoidosis, which was confirmed by transbronchial biopsy.

A high index of suspicion for CS is required as these patients frequently present initially to a cardiologist, and treatment may be focused only towards managing arrhythmias. An accurate and early diagnosis is essential as steroids are also required in such condition.

Our patient was placed on steroid treatment and, although his general condition improved rapidly, he suddenly died one month later.

Bibliography

1. Dubrey S.W., Falk R.H.: Diagnosis and Management of Cardiac Sarcoidosis. *Progr Cardiovasc Dis*, 2010, 52: 336-346.
2. Hunold P., Schlosser T., Vogt F.M., et al.: Myocardial late enhancement in contrast-enhanced cardiac MRI: distinction between infarction scar and non-infarction-related disease. *AJR Am J Roentgenol*, 2005, 184: 1420-1426.
3. Koyama T., Ueda H., Togashi K., et al.: Radiologic Manifestations of Sarcoidosis in Various Organs. *Radiographics* 2004;24:87-104.
4. Morgenthau A.S. and Iannuzzi M.C.: Recent Advances in Sarcoidosis. *Chest*, 2011, 139: 174-182.