Heart failure in a patient with multi-organ sarcoidosis

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Case report

A 53-year-old woman was admitted to our emergency department with progressive shortness of breath during exertion for three months. She had been well before without any remarkable diseases in her medical history and she had never been on regular medication. Physical examination revealed leg oedema, jugular venous congestion and bilateral inspiratory crackles on auscultation of the lungs consistent with biventricular congestive heart failure of unknown aetiology. BNP was markedly increased at a level of 2,812 ng/l. An ECG showed sinus-tachycardia but otherwise was normal.

Echocardiography (fig. 1) revealed dilated cardiomyopathy with severely reduced left ventricular ejection fraction of 20% and diffuse wall motion abnormalities. There was secondary mitral regurgitation stage II. End-systolic pulmonary pressure was elevated (sPAP 50 mm Hg). A chest radiograph (fig. 2A) showed cardiomegaly and diffuse reticulonodular opacifications with patchy consolidation in the right upper lobe and left mid zone. The right hilum was retracted. These findings were confirmed by computer tomography of the chest (fig. 2B), which also revealed mediastinal lymphadenopathy and extensive sub-pleural micro-nodularity, most prominent in the mid and lower lung zones. Moreover, traction bronchiectases in the apices were noted, particularly in the right upper lobe with associated retractile cavitations. Involvement of the pleura was suggested by pleural effusion. The heart was enlarged and surrounded by pericardial effusion. Based on these findings a diagnosis of pulmonic sarcoidosis stage IV with cardiac involvement was suggested. Bronchial biopsy was performed to affirm the diagnosis. Histology revealed multiple non-caseating granulomas consistent with pulmonic sarcoidosis (fig. 3). Cardiac MRI (fig. 4) showed an impaired global cardiac function and reduced anterolateral, septal and apical myocardial contractibility as well as multiple stippled areas of delayed contrast enhancement, particularly in regions of impaired contractility, consistent with myocardial sarcoidosis. Additional granulomatous lesions were found in peripheral bones. Treatment was started with initial high dose steroids as well as a standard heart failure therapy. On the 6-month follow-up visit, the patient was asymptomatic and echocardiography revealed normalisation of the left ventricular dimensions and systolic function.

This case is a striking example of multi-organ involvement of sarcoidosis with heart failure symptoms due to cardiac involvement leading to the initial presentation. In our case, the typical imaging features of mediastinal lymphadenopathy, perilymphatic micro-

Figure 1
Parasternal long axis view showing a dilted left ventricle (LV) and atrium (LA), small pericardial effusion (PE) and pleural effusion (PLE).

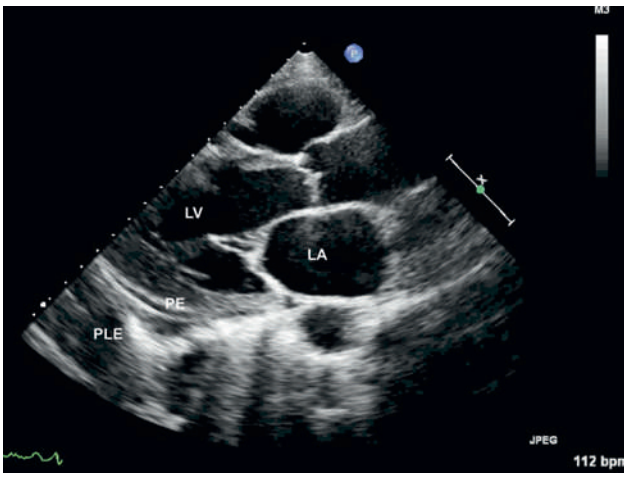

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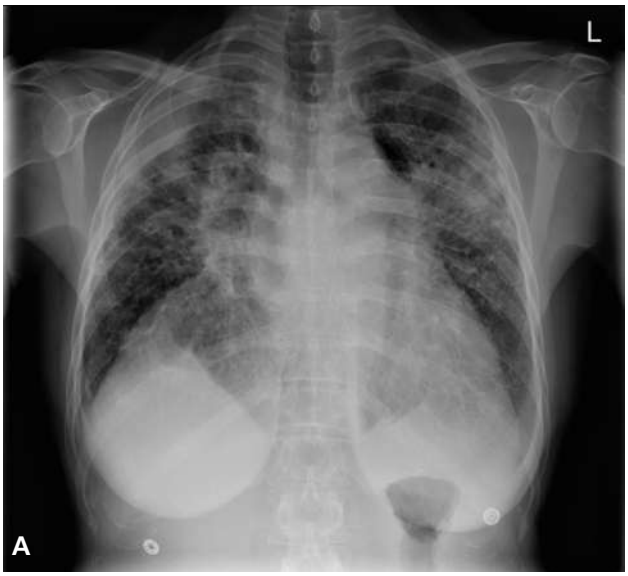
Figure 2
Pulmonary findings.
A Chest x-ray with cardiomegaly and a diffuse reticulonodular opacifications as well as patchy consolidations in the right upper lobe and left mid zone.
B Chest CT (lung window) confirming extensive sub-pleural micronodularity (large arrow), bronchiectasis and scarring in the apices with associated retractile cavitations (small arrow).



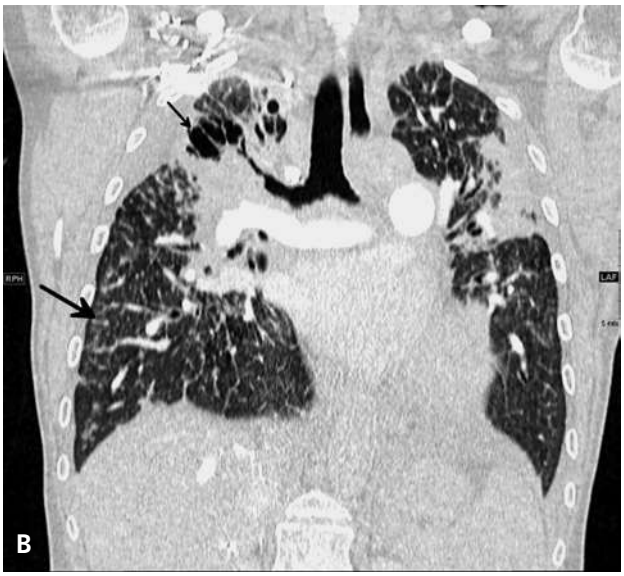


Figure 3
Histologic image of non-caseating granulomas (arrow).

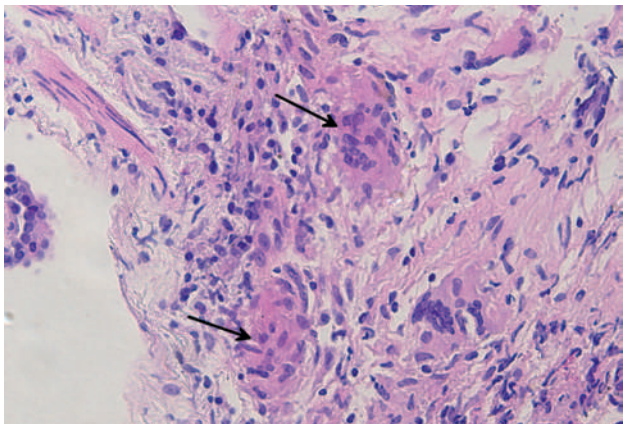
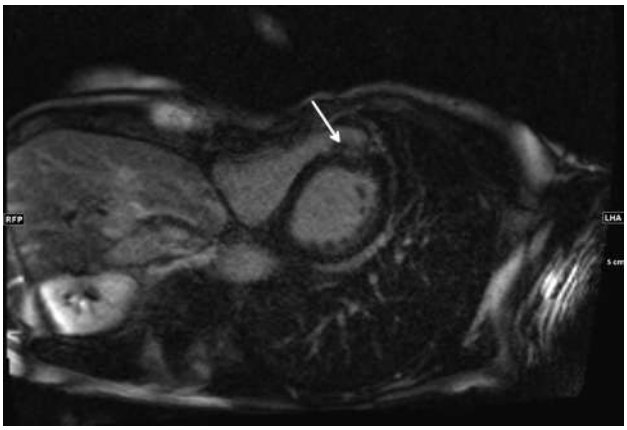


Figure 4
Cardiac MRI in short axis view revealing stippled areas of late contrast enhancement with non-ischaemic pattern (arrow shows contrast enhancement in the anterior wall segment).



nodules and upper lobe predominance alongside typical cardiac manifestations pointed to the diagnosis [1]. Clinical manifestations of cardiac involvement are observed in about 5% of all sarcoidosis patients, although in biopsy studies a cardiac infiltration can be found in up to 25% [2]. When present, congestive heart failure with dilated cardiomyopathy, conduction abnormalities, tachyarrhythmias and even sudden cardiac death are the most common clinical manifestations [2]. Whereas these findings by itself are not indicative of cardiac infiltration of sarcoidosis, concomitant pulmonary involvement strongly suggests the diagnosis.

References

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