CASE REPORT

A patient with a red eye and pulmonary oedema

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SUMMARY

A middle-aged black male patient presented with symptoms and radiological features indicative of pulmonary oedema. Following several admissions for the same symptomatology, and poor resolution of chest radiographic features, the patient developed a red eye. The latter was diagnosed as uveitis, which prompted consideration and proof of a diagnosis of cardiopulmonary sarcoidosis. The patient was subsequently treated with high dose steroids resulting in a partial recovery, complicated by issues of fluid retention.

BACKGROUND

This case illustrates how easy it is to focus on a single diagnosis, in this case left ventricular failure, without an appreciation of the underlying cause. Sarcoidosis remains difficult to diagnose, with no biological marker for the disease. Tissue biopsy confirmation is normally required and is again problematic when it involves the heart due to patchy involvement. This case illustrates a number of procedures ultimately used to confirm the diagnosis.

CASE PRESENTATION

A 52-year-old Nigerian man presented with worsening shortness of breath of 12-month duration. His history included hypertension and atrial fibrillation.

Examination revealed an obese male patient of 142 kg (body mass index 38.6 kg/m²), with blood pressure 150/90 mm Hg and moderate bilateral peripheral oedema to the mid thighs. Blood tests, performed on the fourth and latest admission (12 months after the initial presentation), revealed unremarkable inflammatory markers (C reactive protein 27 mg/L), normal thyroid function, corrected calcium (2.44 mmol/L), serum ACE (50 U/L, normal range 20–90) and troponin I (39 ng/L). The cardiac rhythm had recently deteriorated into atrial flutter at a controlled ventricular rate.

A chest radiograph suggested pulmonary oedema as the cause (figure 1). Echocardiography revealed reasonable systolic function (ejection fraction of 55%), moderate left ventricular hypertrophy (mean 15 mm) and features consistent with diastolic heart failure. Treatment comprised high dose intravenous diuretics and conventional heart failure therapy. Our patient had repeated hospital admissions for shortness of breath with chest radiographs consistently indicative of pulmonary oedema.

Despite sufficient clinical improvement to allow discharge from hospital, radiological improvement was minimal. On the fourth admission in

12 months, the patient developed an acute unilateral red eye. Diagnosed as uveitis, this was treated with topical steroid drops. A brief run of non-sustained ventricular tachycardia prompted a cardiac MR scan.

INVESTIGATIONS

A diagnostic tap of a left pleural effusion revealed an exudate (54 g/L). High resolution CT of the chest (figure 2) showed extensive central mediastinal, paratracheal and perivascular lymphadenopathy, highly suggestive of sarcoidosis. The heart was enlarged with a large left pleural effusion and scattered bilateral pulmonary nodules (largest 1 cm in diameter).

MRI of the heart confirmed left ventricular hypertrophy, with patchy delayed gadolinium enhancement throughout the septum and severe biatrial dilation. Trivial pleural and pericardial effusions along with extensive mediastinal and hilar lymphadenopathy were noted and felt highly compatible with a diagnosis of sarcoidosis.

Bronchoscopy demonstrated diffuse cobblestone appearances. Biopsies of bronchial mucosa showed a stroma infiltrated with lymphocytes and histiocytes with non-caseating granulomas. Staining for fungi and mycobacteria was negative.

DIFFERENTIAL DIAGNOSIS

Hypertensive heart disease, ischaemic cardiomyopathy, idiopathic restrictive cardiomyopathy and hypertrophic cardiomyopathy would be other diagnoses to consider in this patient.

TREATMENT

Intravenous pulsed methylprednisolone for 3 days was followed by oral prednisolone at 40 mg daily thereafter.

Figure 1 Posteroanterior chest radiograph, highlighting cardiomegaly with bilateral perihilar shadowing.

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Figure 2 High resolution CT image slices at two levels (A and B) highlighting pulmonary nodules (1), bilateral hilar lymphadenopathy (2), left-sided pleural effusion (3), thickening of the fissure (4), pulmonary consolidation and cardiomegaly.

OUTCOME AND FOLLOW-UP

Radiological improvement was evident by day 21 of treatment, although the patient subsequently developed considerable fluid retention peripherally. A 25.4 kg gain in weight required further hospital admission to manage this deterioration using intravenous diuretics. Consideration is being given to the implantation of an automated implantable cardioverter-defibrillator (AICD) and to attempting ablation of the established atrial flutter.

DISCUSSION

Estimates suggest that the lifetime risk of developing sarcoidosis among African-Americans may be as high as 2%. Sarcoid involvement of the heart, as with pulmonary disease, may be occult in as many as 50% of cases.¹ Most patients with sarcoidosis are asymptomatic, but the potential for ventricular rhythm disturbance, heart block or cardiac failure has serious consequences. While the incidence for clinical cardiac sarcoid ranges between 4% and 5%,² histological confirmation at autopsy is five times higher. However, confirmation by endomyocardial biopsy is recognised to be unreliable due to the patchy nature of the disease and is not without risk.³

Antiarrhythmic medication,⁴ as well as interventional management with implantable defibrillators, can improve the prognosis, with mortality now skewed towards heart failure, rather than arrhythmias.⁵ An early diagnosis may reduce the infiltrative burden and even halt any progression of heart failure. Heart and other critical organ involvement is usually treated with high dose steroids in the first instance.

The accumulation of fluid in our patient with high dose steroid therapy highlights the issues in treating cardiac sarcoid with such drugs. Recent studies and reviews continue to be sceptical about the long-term use of steroids, or alternative immunosuppressive therapies, in the management of sarcoid disease.⁶ The fluid retention in our patient on receiving steroid therapy is indicative of the severe side effects associated with the immunosuppressive and cytotoxic therapies currently available.⁷

Despite 130 years of investigation, sarcoidosis remains a conundrum, difficult to diagnose with no biological marker and no established aetiology. We present a case of cardiopulmonary sarcoidosis, misdiagnosed as diastolic heart failure for over a year.

The need for clinical acumen remains paramount for this enigmatic but potentially manageable condition.

Learning points

- ▶ The racial group, high body mass index, hypertension and consequent ventricular hypertrophy, delayed identification of the underlying disease process.
- ▶ Afro-Caribbean patients have a higher prevalence of sarcoidosis and a worse prognosis than Caucasians.⁸
- ▶ Hypercalcaemia and serum ACE levels are frequently elevated in patients with severe parenchymal sarcoid lung disease,⁹ but were normal in our patient.
- ▶ Heart failure should prompt a search for an underlying aetiology.
- ▶ Immunosuppressive therapy for sarcoidosis remains a contentious and unresolved issue.^{6, 7}

Competing interests None.

Patient consent Obtained.

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