CASE REPORT

What's wrong with this artery? A medical disease discovered by a surgeon

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Accepted 9 September 2014

SUMMARY

A 50-year-old man presenting with chest pain had positive stress echocardiography; and angiogram showed single artery coronary stenosis, presumed to be atherosclerotic. He was started on optimal medical therapy with good compliance. Four months later, he had a myocardial infarction (MI) and cardiac catheterisation surprisingly showed interval development of severe three-vessel stenosis. He underwent coronary artery bypass grafting (CABG), during which the cardiothoracic surgeon noticed severely inflamed coronary arteries, concerning for vasculitis. Following CABG, the patient continued to have chest pain and was admitted again for MI within 4 months of surgery. Subsequent autoimmune workup was consistent with sarcoidosis. He was started on immunosuppressive therapy for presumed sarcoid-related coronary vasculitis, and 23 months later, the patient has not developed further ischaemic events. This is a rare case and extends the clinical spectrum of cardiac sarcoidosis, presenting with rapidly progressive coronary stenosis most likely due to vasculitis, mimicking atherosclerotic coronary artery disease.

BACKGROUND

Sarcoidosis is a multisystem, inflammatory disease of unknown aetiology, characterised by non-caseating granulomas that may involve any organ in the body. The most common organs involved in sarcoidosis are lungs and lymph nodes. Cardiac involvement was demonstrated in an autopsy study in about 20–30% of patients with sarcoidosis.^{1 2} However, only a fraction of these patients, about 5% or less, presented with symptoms of cardiac involvement during their lifetime.^{1 3–6} The most commonly recognised manifestations of cardiac sarcoidosis include those of conduction abnormalities, congestive heart failure, cardiomyopathy and sudden cardiac death. Epicardial coronary artery infiltration with coronary sarcoid is a recently recognised entity, and is commonly missed as it simulates acute coronary syndrome caused by atherosclerotic cardiovascular disease. We report this unique rare case of cardiac sarcoidosis, presenting with rapidly progressive coronary stenosis most likely secondary to vasculitis.

CASE PRESENTATION

A 50-year-old African-American man with hypertension, diabetes mellitus and peptic ulcer disease, presented to primary care's office in October 2011 with intermittent substernal chest pain of a few months duration. No family history of premature coronary artery disease, and no personal history of

smoking, alcohol or drug misuse. He was referred for stress echocardiographic test, which showed focal wall motion abnormalities and a subsequent coronary angiogram in November 2011, showed 50% stenosis in the proximal left circumflex artery with complete distal occlusion, and normal ejection fraction (EF) of 60% (figures 1 and 2). He was started on optimal medical therapy with aspirin, statin, β -blocker and long-acting nitrate, along with aggressive risk factor modification. Despite excellent compliance, he continued to experience episodes of chest pain but did not seek medical attention for it. In March 2012, 4 months after the first coronary angiogram, he presented to the emergency department with an episode of severe chest pain, with significant changes on ECG (figure 3) and mildly elevated troponin. He was admitted to the hospital with a diagnosis of non-ST-segment elevation myocardial infarction (NSTEMI). He underwent an urgent cardiac catheterisation that surprisingly showed severe interval development of three-vessel stenosis: 95% stenosis in mid and distal left anterior descending (LAD) artery, 90% stenoses in first diagonal branch, left circumflex and obtuse marginal branches and distal right coronary artery (RCA; figures 4 and 5). Left ventricular systolic function was still preserved at 60%. This demonstrated a very rapid progression of coronary stenosis, presumed to be of atherosclerotic origin, in just a brief period of 4 months. He was immediately referred for coronary artery bypass grafting

Figure 1 Cardiac catheterisation-left system, in November 2011 showing 50% stenosis in proximal left circumflex artery with complete distal occlusion.

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To cite: Singh V, Luthra S, Kouides R, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2014-205645

Figure 2 Cardiac catheterisation-right system, in November 2011 showing normal right coronary artery.

Figure 4 Cardiac catheterisation-left system, in March 2012 showing severe three-vessel coronary stenosis.

(CABG), and during the procedure, the cardiothoracic surgeon noticed the coronary arteries did not have a typical atherosclerotic picture, but instead looked severely inflamed, very concerning for some sort of arteritis. Although the aorta looked normal, he did a punch biopsy of it, which later came back normal. Clopidogrel was added to his cardiac regimen after a multivessel CABG. The patient was referred to rheumatology on discharge, but before he could see them, got readmitted in July 2012, for chest pain, with positive ECG changes and troponin levels and was diagnosed with NSTEMI. He mentioned still having intermittent chest pain post-CABG, but chose to ignore them for a while. This time, a third angiogram within the past

9 months showed three-vessel coronary stenosis as before, with widely patent 5 of 6 bypass grafts and overall preserved left ventricular systolic function. Cardiology recommended adding ranolazine to his optimal medical regimen. Rheumatology was consulted in-house now, for the high suspicion of a vasculitis and a detailed autoimmune workup was positive for a high titre of nucleolar pattern antinuclear antibodies and high levels of erythrocyte sedimentation rate, C reactive protein and ACE. Hepatitis panel, HIV screen, antimyeloperoxidase and antiproteinase antibodies were negative. He was started on empiric high-dose corticosteroids. CT chest showed bilateral mediastinal and hilar adenopathy, with multiple pulmonary nodules

Figure 3 ECG, in March 2012 showing normal sinus rhythm, rate of 88, with T wave inversions in leads V4 through V6 and mildly depressed in leads I and aVL (arrows).

Figure 5 Cardiac catheterisation-right system, in March 2012 showing severe three-vessel coronary stenosis.

(figure 6). Mediastinoscopy and biopsy of mediastinal nodules showed non-caseating granulomatous highly consistent with sarcoidosis. Cardiac MRI showed left ventricular basal inferolateral and mesocardial enhancement again pointing towards an infiltrative process suggestive of sarcoidosis (figure 7).

TREATMENT

He was discharged from the hospital on high-dose oral prednisone (60 mg oral daily), which was tapered to a low maintenance dose (15 mg oral daily) after the addition of methotrexate (20 mg oral weekly) and hydroxychloroquine (200 mg oral twice a day).

OUTCOME AND FOLLOW-UP

He has been on immunosuppressive therapy for coronary sarcoidosis for 23 months, and as of now, has not had any further hospital admissions for ischaemic events.

DISCUSSION

We found only three case reports that are reporting coronary involvement related to sarcoidosis. One case report described a 47-year-old Caucasian man with isolated cardiac sarcoidosis in the form of non-necrotising granulomas in the walls of

Figure 6 CT scan of chest with contrast, in July 2012 showing bilateral mediastinal and hilar adenopathy (arrows), with multiple pulmonary nodules.

Figure 7 Cardiac MRI from July 2012, short axis view showing late basal inferolateral wall enhancement (arrow).

epicardial coronary arteries, found on histological examination of his explanted heart after he underwent an orthotopic heart transplant for presumed idiopathic dilated cardiomyopathy.⁷ This patient presented with progressive congestive heart failure despite aggressive medical management. His two-dimensional echocardiogram showed a reduced EF and normal coronary anatomy on angiogram. In another case report of a 40-year-old Caucasian man who presented with increasing shortness of breath and chest pain and was diagnosed with a complete heart block. Cardiac MRI demonstrated epicardial fat infiltration surrounding the large coronary arteries. Echocardiography showed an EF of 30% and cardiac catheterisation showed normal coronaries. His ACE levels were high and biopsy of a subcutaneous lesion on the torso showed non-caseating granulomas confirming the diagnosis of sarcoidosis.⁸

In both these cases, angiogram was normal and EF was low. Notably, in our case the EF remained preserved, despite such a diffusely diseased coronary vasculature demonstrating that this was not primarily a myocardial process.

We found only one case report of biopsy proven coronary sarcoidosis of a 68-year-old African-American man, with a known history of non-caseating granulomas found on lymph-node biopsy who was on the treatment with Prednisone.⁹ He presented with a MI, 6 years following his diagnosis with sarcoidosis and underwent stenting of the LAD. He presented with restenosis of the culprit vessel four times over the following year despite target vessel revascularisation at each presentation. Biopsy from the epicardium of the RCA revealed non-caseating granulomatous inflammation. He was treated with prednisone and mycophenolate mofetil. However, in this case report, the patient had an established history of sarcoidosis prior to his coronary disease, in contrast to our patient where the diagnosis of sarcoidosis was performed with coronary vasculitis as the initial presentation.

Cardiac sarcoid typically presents as arrhythmias, conduction defects and heart failure. This case extends this spectrum of clinical presentations as it could present with progressive vasculitic coronary stenosis that mimicks coronary artery disease of atherosclerotic origin. In summary, our patient presented with rapidly progressive stenotic ischaemic heart disease that

continued to present with ischaemia despite a CABG. He was diagnosed with lymph node proven sarcoidosis, had evidence of infiltrative disease in the myocardium and eventually had clinical response to immunosuppressive treatment.

Learning points

- ▶ Cardiac sarcoidosis is rare, and concomitant involvement of the epicardial coronary arteries is even more rare.
- ▶ Cardiac sarcoidosis typically presents as arrhythmias, conduction defects and heart failure.
- ▶ This case extends this spectrum of clinical presentations of cardiac sarcoid as it could present with rapidly progressive vasculitic coronary stenosis most mimicking coronary artery disease of atherosclerotic origin.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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