Rare disease

Rare manifestation of Behcet's syndrome: insight from multimodality cardiovascular imaging

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Summary

A young Jordanian man was hospitalised with fever of 2 months duration. Preadmission diagnostic assessment including CT thorax and abdomen were inconclusive. Right-sided pleuritic chest pain was present on admission. Pulmonary embolism was suspected on a ventilation/perfusion lung scan. Lower-extremity deep venous thrombosis was, however, absent. Echocardiogram to exclude a cardiac source of pulmonary embolism showed a right ventricle mass which was also present on retrospective review of prehospitalisation CT thorax. Tissue characterisation of this mass on cardiac MRI was not helpful. Empiric anticoagulation was started without reduction in size at 3 weeks. Due to ongoing diagnostic uncertainty, surgical resection was performed. Histopathology confirmed this mass to be a thrombus. With unabated fever, right ventricular thrombosis with pulmonary embolism and cutaneous and scrotal ulceration which evolved later in the hospital course, a diagnosis of Behcet's syndrome was considered and subsequently confirmed. Response to immunosuppressive therapy was prompt.

BACKGROUND

Behcet's syndrome (BS) is a multisystem vasculitis with a higher prevalence in the Mediterranean region. Cardiovascular complications of BS may precede other well-recognised manifestation of mucocutaneous ulceration and uveitis. In reported clinical series of BS cardiac involvement is seen in only 1–5% of patients.¹ Cardiac thrombi, which usually affect the right side of the heart, are rarer still. However, cardiac involvement is more frequently observed on autopsy.² Early recognition of cardiac involvement in BS can influence the diagnostic course and therapeutic outcome. It is largely dependent on cardiac imaging that should be considered in all known and suspected cases of BS.

CASE PRESENTATION

An 18-year-old Jordanian man was hospitalised with intermittent fever of 2 months duration. Diagnostic studies prior to his admission were unrevealing. Patient was earlier placed on colchicine for a provisional diagnosis of Familial Mediterranean Fever. There was, however, no measurable improvement on treatment. Other than intermittent fever which responded to acetaminophen, there were no signs of mucocutaneous inflammation or ulceration, arthralgia, cough or sputum expectoration or bladder or bowel disturbance.

The patient had no significant medical history. There was no history of travel abroad. He was a high-school student at the time, was single and sexually inactive. Family history was non-contributory to his care. Medications included colchicine and acetaminophen.

Physical examination showed a young man of average height and built and in no obvious distress. He had a low-grade fever. Vital signs were otherwise normal. There was

no conjunctival injection, skin or mucosal ulceration, lymphadenopathy or joint inflammation. Cardiovascular, respiratory, neurological and abdominal examinations were also normal.

Acute phase reactants including erythrocyte sedimentation rate and C reactive protein were elevated. There was no leucocytosis, renal function and liver function tests were normal. Repeated blood cultures, malaria smear and urine culture remained negative. Hepatitis serology was negative and titres of antinuclear antibody and antineutrophil cytoplasmic antibody were not elevated. Preadmission contrast CT chest and abdomen as work-up of fever of unknown origin was reported as normal with no evidence of lymphadenopathy or hepatosplenomegaly.

For symptoms of pleuritic right-sided chest pain on admission, a ventilation/perfusion lung scan was performed which showed perfusion mismatch in the distribution of right pulmonary artery consistent with a diagnosis of pulmonary embolism. Therapeutic anticoagulation was started. Lower-extremities duplex ultrasound was negative for deep vein thrombosis. Screening for thrombophilic disorders which included protein C and S, anticardiolipin antibody and factor V Leiden, which were requested prior to the start of anticoagulation, was unrevealing.

Echocardiography was subsequently performed to exclude right-sided cardioembolic cause as well as to assess the impact of pulmonary embolism on right ventricular function. There was no evidence of endocarditis on echo. Right ventricular cavity was not dilated and overall right ventricular as well as left ventricular function was normal. Incidentally, a mass protruding from the septal wall of the right ventricle was seen on echo. Echo appearance was inconclusive as was tissue characterisation on cardiac MRI. Differential diagnosis of this mass included cardiac tumour,

possible vegetation on the endocardial surface of the right ventricle or a mural thrombus. Of interest, on retrospective review of prehospitalisation contrast CT chest, this mass was also present (see online supplementary figure S1). In view of the possibility of a cardio thrombo-embolic disorder a trial of therapeutic anticoagulation was decided upon. However, no reduction in the size of this mass was noted on follow-up echocardiogram 3 weeks later. With no apparent clinical or echocardiographic improvement and remaining diagnostic uncertainty, surgical excision of the right ventricular mass was performed. Histopathology confirmed this to be a thrombus. This latter finding combined with unabated fever and cutaneous and scrotal ulceration that evolved later in the hospital course a diagnosis of BS was considered and subsequently confirmed with a positive cutaneous pathology test.

The patient was placed on high-dose oral steroids and immunosuppressive therapy with azathioprine. Anticoagulation was maintained. Resolution of fever was prompt, followed by healing of skin and scrotal ulcers. The patient has remained asymptomatic at 1-year follow-up.

DISCUSSION

The classic triad of recurrent oral and genital ulceration with uveitis in BS may be preceded by non-specific multisystem vasculitis affecting both the arterial and venous system.¹ Vascular involvement in particular deep vein thrombosis and thrombophlebitis is more common in men.^{1 3} Despite high incidence of vascular damage, cardiac involvement in clinical reports of BS is uncommon varying from 1% to 5%.¹ However, in an autopsy series of BS from Japan higher cardiac involvement was reported.² Ranging from coronary involvement to pancarditis and cardiomyopathy, cardiac BS can be a diagnostic and therapeutic challenge.² In very rare cases intracardiac thrombus formation which favours the right side of the heart is noted.^{4 5} Such cases are associated with a higher occurrence of lower-extremity deep vein thrombosis and thrombosis of inferior vena cava.^{6 7}

Diagnostic use of contrast-enhanced chest and abdominal CT is not uncommon in patients with fever of unknown origin. Although, echocardiography is the preferred imaging modality for detection of cardiac thrombi, it is possible to suspect or detect cardiac masses on contrast-enhanced chest CT, particularly those performed with ECG gating. Uniform contrast opacification of the cardiac chambers is, however, necessary to avoid artefact created by contrast streaming in cardiac chambers. Additionally, in cases of suspected cardiac mass or a mass of unclear aetiology cardiac MRI can be used for diagnostic confirmation with the added advantage of tissue characterisation. In our opinion routine echocardiography should be considered in all patients with BS for early detection of potentially life-threatening cardiac and pulmonary complications. Cardiac CT and MRI can be reserved for patients with limited echo windows, where there is remaining uncertainty about cardiac involvement and in those where additional information regarding coronary anatomy and myocardial involvement is considered important. In our

patient retrospective review of chest CT clearly demonstrated a right ventricular mass (see online supplementary figure S1) as was later confirmed on echo. Earlier recognition of this cardiac mass may have accelerated the diagnosis of BS and avoided a potentially life-threatening complication of pulmonary embolism. Why cardiac MRI was not helpful in establishing the nature of this mass may have to do with technical factors and our limited institutional expertise in the use of cardiac MRI.

Management of intracardiac thrombus in BS is controversial.⁵ Medical management is preferred as a first-line intervention^{4 6 7 8} with surgical resection reserved in cases of failed medical therapy, recurrent or large cardiac thrombus and where there is diagnostic uncertainty.⁹

Learning points

- Cardiac thrombus though uncommon and perhaps in some cases unrecognised may be an early manifestation of Behcet's syndrome (BS) preceding other well-known clinical features of BS.
- In appropriate cases, BS should be considered in cases of thromboembolic disorders where an underlying cause is not apparent. This is perhaps more relevant in patients of Mediterranean origin given the disease prevalence.
- Conservative management with anticoagulation and immunosuppressants are usually effective with resolution of cardiac thrombi in the vast majority. Surgical extraction of cardiac mass may be considered in cases where the diagnosis is uncertain or when medical therapy in thrombus resolution is unsuccessful.

Competing interests None.

Patient consent Obtained.

► An additional supplementary figure is published online only. To view this file please visit the journal online (<http://dx.doi.org/10.1136/bcr-2012-007148>)

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