Acute Congestive Heart Failure Secondary to an Unusual Underlying Cause

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Abstract

We report a patient who presented with non-specific features and rapidly developed multisystem disease as a result of Churg-Strauss syndrome, a rare diffuse primary vasculitis. This case report highlights the importance of considering primary vasculitides as a differential diagnosis in patients presenting with multiple organ involvement as early specific therapy in such cases has shown to change the outcome. ©

INTRODUCTION

In patients presenting with multisystem disease, vasculitis should always enter into the differential diagnosis. Whilst blood tests may be helpful in reaching a diagnosis, the importance of new and apparently specific tests such as anti-neutrophil cytoplasmic antibodies can be overestimated at the expense of traditional assessments such as a good history and comprehensive clinical examination and simple tests such as a blood count. We report a patient who presented with non-specific features and rapidly developed multisystem disease and severe congestive heart failure refractory to conventional treatment. Diagnosis of primary vasculitis in this case led to definitive treatment with excellent recovery.

CASE REPORT

A 61-year man was admitted initially under the surgeons with three-week history of intermittent upper abdominal pain, nausea and anorexia. Five years ago he was diagnosed to have late onset asthma and over the past three years had recurrent episodes of nasal trouble diagnosed as sinus inflammation by ENT surgeons. Eight weeks prior to admission his peak flow dropped what he felt was an exacerbation of asthma and was prescribed a 5-day course of 30 mg Prednisolone. On examination he was noted to have epigastric and right upper quadrant tenderness. Apart from normocytic normochromic anaemia (Hb 11.3g/dL), leukocytosis with eosinophilia (Eosinophils 4.6 x 10^9 /L, 36 % of total leukocyte count) and elevated CRP (49 mg/L) his initial blood tests were normal. Chest X-ray (CXR) demonstrated non-specific shadowing in upper zones. Abdominal ultrasound and CT scan demonstrated bilateral pleural effusions and a small pericardial effusion. One week after admission he had developed persistent swinging pyrexia (range 38-39°C) and was commenced on broad-spectrum antibiotics for presumed atypical pneumonia.

After three days he became very dyspnoeic and developed significant peripheral oedema with a raised JVP and widespread lung crepitations. ECG demonstrated non-specific ST-T changes; CXR demonstrated cardiomegaly and widespread ground glass shadowing and both Troponin-I (1.29mg,normal <0.10 mg) and B-type natriuretic peptide (286 ng/L, in heart failure >100 ng/L) were elevated. He was commenced on intravenous diuretic and nitrate therapy along with low molecular weight heparin and fluid restriction with a diagnosis of severe congestive heart failure (CHF) secondary to an acute coronary event. After 4 days symptoms related to CHF had improved only marginally (at this stage an echocardiogram demonstrated pericardial effusion and left ventricular ejection fraction reduced at 42%). His pyrexia remained unabated with persistently raised inflammatory markers and marked eosinophilia. His repeated blood and urine cultures did not grow any organism and atypical pneumonia screen was negative.

He then developed palpable purpura spread over his thighs and buttocks and this prompted a rheumatological referral. A unifying diagnosis of Churg-Strauss syndrome (CSS) was made based on Lanham’s criteria (asthma, peak peripheral eosinophil count >1.5x 10^9 /L, 36 % of total leukocyte count) and elevated CRP (49 mg/L) his initial blood tests were normal. Chest X-ray (CXR) demonstrated non-specific shadowing in upper zones. Abdominal ultrasound and CT scan demonstrated bilateral pleural effusions and a small pericardial effusion. One week after admission he had developed persistent swinging pyrexia (range 38-39°C) and was commenced on broad-spectrum antibiotics for presumed atypical pneumonia.

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and became apyrexial. By 4th day both his inflammatory markers and eosinophil count had settled down to the normal ranges and he was well enough to be discharged home. Follow up has shown that he has remained well since with a decreasing dose of steroid.

DISCUSSION

This man with abdominal pain had a clear history of late onset asthma and paranasal sinus symptoms, had eosinophilia and developed lung involvement, vasculitic rash and heart failure refractory to conventional treatment.

The 1994 Chapel Hill consensus conference defined Churg-Strauss syndrome as an “eosinophil-rich granulomatous inflammation of the respiratory tract, necrotizing vasculitis affecting small to medium sized vessels and associated with asthma and eosinophilia”. Diagnosis is based mainly on clinical grounds since eosinophilic granulomas are not always present on biopsy even in florid cases. Lanham’s clinical criteria are widely used, and he described three phases of the condition: firstly, an allergic rhinitis, often associated with recurrent sinusitis and asthma, that becomes progressively more difficult to treat; secondly, peripheral blood eosinophilia associated with eosinophilic pulmonary infiltrates and worsening asthma; and thirdly, systemic vasculitis, commonly including peripheral neuropathies and occasionally life threatening cardiac and severe gastrointestinal disease.

The spectrum of cardiac involvement ranges from pericarditis and pericardial effusion and to more serious cardiac tamponade, CHF caused by an eosinophilic myocarditis and coronary vessel vasculitis. In our patient abdominal pain and elevated Troponin-I can be explained by mesenteric vasculitis and coronary vessel vasculitis respectively. Angiography of relevant vasculature in acute phases may demonstrate multiple vessel wall irregularities, abrupt terminations and small aneurysms. Delay in recognising the cardiac involvement probably accounts for the high cardiac mortality in CSS.

Whilst eosinophilia is a diagnostic marker, antineutrophil cytoplasmic antibodies (ANCA) are present in only about half of patients, with a perinuclear staining and the demonstration of anti-myeloperoxidase antibodies. CSS often responds rapidly to glucocorticoids alone. Cyclophosphamide may be used as an adjunct therapy in patients who have substantial vasculitic end organ involvement and in those who have not proven responsive to the glucocorticoids.

We draw the attention of physicians providing acute medical care to the continuing importance of vasculitis in the differential diagnosis of multi-system disease.

REFERENCES