

An Interesting Case of Pyrexia of Unknown Origin

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Abstract: Pyrexia of unknown origin (PUO) is one of the major diagnostic challenges. A 43 year old patient with rheumatic heart disease (RHD) presented with PUO and suspected to have subacute bacterial endocarditis (SBE) was treated only to be later diagnosed with connective tissue disorder Systemic Lupus Erythematosus (SLE) as the cause. Two disease entities occurring with similar presentations pose challenges both diagnostically and therapeutically making it essential for physicians to have a high index of suspicion and initiate treatment.

Keywords: Pyrexia of unknown origin (PUO), Systemic Lupus Erythematosus (SLE), Rheumatic heart disease (RHD)

INTRODUCTION

Pyrexia of unknown origin (PUO) refers to prolonged febrile illness that persists without diagnosis after careful initial assessment. Although over 200 causes have been described including rare diseases most cases are due to familiar entities presenting in an atypical fashion [1]. Here we are reporting a case of PUO in A 43 year old patient with rheumatic heart disease.

CASE REPORT

A 43-year-old female, house-wife, presented to our hospital with history of fever since 6 months, palpitations since 6 months, low grade fever, continuous, not associated with chills & rigors, evening rise of temperature, not relieved with medication. Palpitations were insidious in onset, progressive, regular, continuous, aggravated on exertion. Patient gave h/o loss of appetite & loss of weight of approximately 16kg since last 6 months, generalised weakness & easy fatigability. No h/o joint pains, cough, decrease urine output, vomiting, loose stools, swelling of lower limbs, headache, chest-pain, abdominal pain, breathlessness. No h/o menstrual abnormalities.

Patient was diagnosed with RHD 22years back & was on regular penicillin prophylaxis treatment and K/C/O hypothyroidism since 10 years & on treatment.

General physical examination revealed that the patient was moderately built & poorly nourished. Malar

rash was present. Pallor & grade 2 clubbing were present, no cyanosis, lymphadenopathy, pedal oedema. Pulse rate was 68 beats/min, regular, rhythmic, high volume, collapsing in nature, all peripheral pulses were felt, pulses bisferiens felt in carotids, no radio-radial /radio-femoral delay. JVP was raised with prominence of 'a' wave. Blood pressure was 100/60 mm of Hg taken in right arm supine posture.

On cardio-vascular system examination – apex beat was found to be in 6th ICS lateral to the mid-clavicular line. On auscultation mitral area- S1 & S2 heard, loud S1, MDM of grade 3/6 & PSM of grade 4/6 were heard. EDM of grade 4/6 is heard in aortic area with soft A2, EDM of grade 3/6 is heard in neo-aortic area.

Abdominal examination revealed hepato-splenomegaly. Other systems were normal.

With the above mentioned history & examination a diagnosis of subacute bacterial endocarditis was suspected & relevant investigations were done.

Complete blood count – anaemia (Hb - 9.7g %) with thrombocytopenia; Urine routine – 250 erythrocytes, no proteinuria/leukocytes; RFT & LFT – Normal; Serum electrolytes were normal; Peripheral smear for malarial parasite- negative; Dengue – negative; ESR – 70 mm/hr; Ultrasound abdomen –

cholelithiasis (10mm); Bilateral renolithiasis (r- 3.8mm, l – 4.1mm); Blood culture – no growth (repeated twice).

Echocardiography - RHD, severe AS, moderate AR, moderate MS, mild MR, LV DILATED, mild LV dysfunction, trivial TR, noPAH, good RVfunction, no clot/vegetation/PE. RVSP-25mm Hg, EF- 45-50%

Patient was treated with IV antibiotics, gentamycin & ceftriaxone for 1 week but fever persisted. Patient was investigated further to rule out connective tissue disorders in the wake of high ESR, multiple oral ulcers, malar rash, anaemia and microscopic haematuria.

CRP – raised, ASO – normal; ANA – 3+; ANA Profile – SSA, Ro-52, SSB, anti-dsDNA were positive.

With the ANA profile patient was diagnosed with Systemic Lupus Erythematosus (SLE). Patient was treated with IV steroids following which her symptoms reduced within 2days & became afebrile after the initiation of steroids.

Patient was treated with oral prednisolone & mycophenolate mofetil at the time of discharge & asked to follow up every 3 months.

DISCUSSION

SLE is an auto-immune rheumatic disease in which organs & cells undergo damage initially mediated by tissue binding auto-antibodies & immune complexes [2].

Diagnostic criteria include -Malar rash, discoid rash, photosensitivity, oral ulcerations, arthritis, serositis, renal disorder, neurologic disorder, hematologic disorder, immunologic disorder, anti-nuclear antibodies [2].

Etiology is multifactorial, incorporating genetic, hormonal & environmental elements, no single abnormality of the immune system can be considered responsible. Pathogenesis depends on the interplay of a number of different factors including auto-antibodies, T-lymphocytes, cytokines, the complement system & apoptosis [3].

Valvular abnormalities can be found on echocardiography in up to 54% of patients with SLE [4]. Most common is non-specific thickening of mitral & aortic valve [5, 6]. Untreated SLE per se may produce severe organic valvular disease [7]. The most characteristic valvular abnormality of SLE is Libman-Sacks endocarditis [8]. Immunoglobulin and complement deposition in the valvular structure will

subsequently lead to Libman-Sacks vegetations, valve thickening, and valve regurgitation [9].

It has been reported that significant morbidity and mortality may result from SLE valvular heart disease in about 1 to 2 percent of SLE patients. The pathogenetic mechanisms of valve dysfunction in SLE patients are reported to be multifactorial [10].

In the current case study blood culture was negative for any growth, echocardiography did not show any vegetations, patient was not responding to IV antibiotics, hence the possibility of infective endocarditis was ruled out and the etiology for PUO was looked for.

CONCLUSION

Subacute bacterial endocarditis and SLE both are causes of PUO and both essentially treatable. In the setting of a new emergent disease in a pre-existing one, identification of them is essential and early institution of treatment is the need for better prognosis and reduction of complications and mortality.

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