

Case Report

OMINOUS DISEASE WITH AN UNUSUAL CLUE: BACTERIAL ENDOCARDITIS PRESENTING AS CUTANEOUS VASCULITIS

*Anuj Gupta, JoshikaAgarwal, Jay Saparia, SamidhTyagi, TejasBhalodia

Independent Researcher, India.

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ABSTRACT

Sub-acute bacterial endocarditis (SABE) has varied manifestations with a few less common presentations like cutaneous vasculitis. We present one such case of a middle aged female who presented with a painless rash involving both lower extremities. She was later found to have vegetations on echocardiogram. She was treated with appropriate antibiotics for bacterial endocarditis. Her rash resolved subsequently in a few days suggesting the etiology of rash as infective endocarditis. Hence, vasculitis rash should prompt both infective and immunological workup combined with skin biopsy to aide in specific diagnosis.

Keywords: vasculitis, endocarditis, rash, infective endocarditis

INTRODUCTION

Sub-acute bacterial endocarditis can have different presentations. Cutaneous vasculitis can be a presenting feature in different medical conditions, explained by the effect of circulating immune complexes and micro emboli on the vascular endothelium, therefore the diagnosis of infective endocarditis may be delayed. In this case report, we present a female patient who initially presented with a picture of cutaneous vasculitis delaying the diagnosis of an underlying infective endocarditis with aortic, mitral and tricuspid valve involvement.

CASE PRESENTATION

A 47 year old female with past medical history of multiple sclerosis and congenital ventricular septal defect was admitted for dyspnea and non-blanching purpuric rash involving bilateral lower extremities. Detailed anamnesis revealed fatigue, weight loss and bilateral lower extremity edema, without fever or other specific symptoms. She was found to be in congestive heart failure. Outpatient medications included beta interferon for multiple sclerosis. On physical examination, grade 3/6 systolic murmurs was heard in the left third intercostal space along the mid-clavicular line, and multiple, palpable purpuric lesions with blistering was noted on both legs. No other relevant findings were elicited.



Figure 1: Palpable purpuric rash in bilateral lower extremity

Blood tests revealed white blood cell count 7.7, red blood cell count 3.18, hemoglobin 7.7 g/dL, platelet count 166 k/cumm, iron 11 mcg/dL, direct iron binding capacity 184 mcg/dL, percentage saturation 6, blood urea nitrogen 24 mg/dL, creatinine 1.42 mg/dL (baseline creatinine 0.69 mg/dL). Of note, ferritin 1077.5 ng/mL, hypocomplementemia with C3 complement 25 and C4 complement 8, creatine kinase 11 L, erythrocyte sedimentation rate (ESR) 63, C reactive protein 84 mg/L, haptoglobin 192 mg/dL, IgA serum 224, IgG

serum 1965, IgM serum 148 and BNP 1022 pg/mL. The following labs were negative: hepatitis panel, anti-nuclear antibody (ANA), Anti-double stranded DNA, anti-neutrophil cytoplasmic antibodies (c-ANCA). Urine analysis showed protein 100 mg/dL, 20-50 red blood cells, 20-50 white blood cells, negative nitrate. Urine protein to urine creatinine ratio 1848. Blood cultures taken grew *Granulicatella adiacens*, family of nutritionally variant streptococci (NVS) Transesophageal echocardiogram demonstrated 1.08 cm vegetations on aortic valve with severe insufficiency, small vegetation on the anterior mitral valve leaflet with moderate mitral regurgitation, moderate tricuspid regurgitation with a small mobile vegetation. Skin biopsy results from bilateral lower extremities reported mixed cell infiltrate with neutrophils and vascular thrombosis with hemorrhage and vascular thrombosis with neutrophils and a trace of karyorrhexis. The two biopsies showed overlapping histological features, and did suggest a septic/thrombotic vasculopathy. The patient remained afebrile throughout the first 7 days of hospitalization. Patient was treated with ceftriaxone and vancomycin for infective endocarditis. The patient's condition improved, and her purpuric rash disappeared.

DISCUSSION

Infective endocarditis is a multisystem disease that can sometimes lead to a diagnostic challenge. The most common extra cardiac manifestations of infective endocarditis that have been reported are splinter hemorrhages, Osler's nodes, Janeway lesion, Bowman lesions of the eye, Roth spots, petechiae, and clubbing. [Silverman and Upshaw, 2007] However, there are a variety of embolic and immunological manifestations that a physician should consider when encountering a patient with palpable purpura in the lower extremities and while diagnosing infective endocarditis. A study reported that 3.5% of the patients presenting with cutaneous vasculitis had an underlying infective etiology; with 0.7% having endocarditis as the underlying cause. [Loricera *et al.*, 2015] Another study done on 172 patients, reported underlying cause of cutaneous vasculitis as infective etiology in 2.9% patients; with 2.3% having endocarditis. [Blanco *et al.*, 1998] Skin biopsy can help differentiate between cutaneous vasculitis associated with severe bacterial infection and cutaneous vasculitis secondary to other causes; with the former showing neutrophilia as seen in our patient presented above. [Loricera *et al.*, 2016] Bacterial endocarditis may sometimes mimic HSP with renal involvement (hematuria and proteinuria) as seen in our patient. This clinical scenario is not uncommon and studies have shown that 41% of patients with infective endocarditis have rheumatological manifestations including vasculitis at initial presentation. [González-Juanatey *et al.*, 2001] Our patient had palpable purpura that are typically seen in classic HSP, [Sohagia *et al.*, 2010] hence it lead to further investigations towards vasculitis. The renal involvement in patients with infective endocarditis can be due to variety of factors with the most common cause being localized infarct secondary to septic emboli. [Majumdar *et al.*, 2000] Though our patient was ANCA negative, multiple studies have reported various pathogens that have been known to mediate ANCA productions in patients with infective endocarditis including *Staphylococcus*, *Streptococcus*, *Tuberculosis bacilli*, *Legionella*, *invasive Aspergillus*, hepatitis C virus, parvovirus B19, and *Bartonella*. [deCorla-Souza and Cunha, 2003; Satake *et al.*, 2011; Chou *et al.*, 2000] In line with the above studies, some studies reported that infective endocarditis with ANCA positivity who were continued to be treated with antibiotics without immunosuppressive therapy had a disappearance of ANCA level and normalization of complement levels. [11] Physicians should rule out sub acute bacterial endocarditis as a probable diagnosis before committing a patient to long-term immunosuppressive therapy on finding ANCA

antibodies in a patients suspected of having systemic vasculitis. [Choi *et al.*, 2000] But in cases where infective endocarditis presents with ANCA positivity, and rapid clinical decline is observed even after appropriate antibiotic treatment, initiating corticosteroids and immunosuppressive therapy is justified and has shown to have clinical improvement. [13,14] There have also been cases reported where systemic vasculitis presented as likely infective endocarditis. [Marco Calachanis *et al.*, 2003]

CONCLUSIONS

Thus it is important to differentiate between infective etiology of vasculitis versus systemic vasculitis given the overlap of the symptoms and the laboratory studies.

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