

CASE REPORT

Right-sided endocarditis in an adult patient with hyperimmunoglobulin-E syndrome successfully treated with high-dose daptomycin

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Abstract

Hyperimmunoglobulin-E syndrome is a syndrome of recurrent staphylococcal abscesses, sinopulmonary infections, and severe eczema, which is characterized by increased IgE and specific facial features. Herein, we describe a patient with hyperimmunoglobulin-E syndrome and right-sided endocarditis due to *Staphylococcus aureus*, which was successfully treated with high-dose daptomycin.

Keywords

Staphylococcus, Tricuspid valve endocarditis, Daptomycin, Hyperimmunoglobulin-E syndrome

1 Introduction

Hyperimmunoglobulin-E syndrome (HIES) is a syndrome of recurrent staphylococcal abscesses, sinopulmonary infections, severe eczema and elevated levels of serum IgE^[1, 2]. Herein, we describe a patient with HIES and right-sided endocarditis due to *Staphylococcus aureus*.

2 Case presentation

A forty-two years old male patient presented to the hospital with fever and rigors of one week's duration. On examination, the patient appeared acutely ill, with dyspnea, edema of both legs, crackles on lung auscultation, a temperature of 40 °C and tachycardia. A transthoracic cardiac triplex, which was performed due to signs of right-heart failure, revealed the presence of a vegetation in the tricuspid valve, with concomitant major tricuspid valve regurgitation (see Figure 1). The patient was admitted to the hospital.

From his past medical history, he was suffering from eczema and had many respiratory tract infections in the past, some of which required admission to the hospital. He denied being an intravenous drug user and he had been working as a manager in a big company.

Five out of five blood cultures yielded *Staphylococcus aureus*, sensitive to methicillin and the presence of right-heart endocarditis was documented. Dicloxacillin 2g every six hours was administered. Due to the presence of crackles on both lungs, a spiral CT was performed, which revealed the presence of multiple septic emboli as well as pleuritic fluid, that was compatible with an exudate (see Figure 2). As the patient remained febrile while receiving oxacillin, the regimen was changed to high-dose daptomycin and rifampin. Vancomycin was not administered to the patient due to impaired renal function, which was attributed to glomerulonephritis. As daptomycin has been documented to be successful in the treatment of right-sided endocarditis, it was chosen instead of vancomycin because it seems not to worsen renal function. With the administration of daptomycin and rifampin for endocarditis, the patient gradually became afebrile and he was discharged from hospital having only mild edema in the legs and no crackles in the lungs.

Due to the presence of right-sided endocarditis, which is much more common in intravenous drug users than in the general population, we began to explore what could have been responsible for the right-sided endocarditis in this patient. Due to its medical history of many respiratory tract infections and eczema, the presence of HIES was suspected, which was confirmed with the finding of serum IgE levels of 1776 UI/L. Thus, the presence of HIES could account for the right-sided endocarditis due to *Staphylococcus aureus* in this patient.



Figure 1. The tricuspid's valve vegetation sizing 18.8 mm ×10.2 mm



Figure 2. Spiral CT scan depicting multiple septic emboli presenting as lung abscesses.

3 Discussion

HIES, which was first described in 1966 and was called Job's syndrome, is characterized by specific facial characteristics, together with the presence of increased IgE levels, eczema and recurrent infections, which are usually caused by *Staphylococcus aureus* [2-5]. The most characteristic facial feature is a broad nasal base and broad nasal bridge, protrusion of the forehead, wide outer canthal distances, and deep set eyes [6, 7].

HIES is due to defects in Janus activated kinase/signal transducer and activator of transcription (JAK-STAT)-mediated cytokine signals, including interleukin (IL)-6 and IL-23 [8-13].

To our knowledge, right-sided endocarditis has only once been reported before in a patient with HIES [14]. It seems likely that *Staphylococcus aureus* found its way to cause bacteremia and tricuspid valve endocarditis through the eczematous skin of the patient. The defective immune response due to the presence of the HIES could account for the presentation with right-sided endocarditis and multiple pulmonary emboli. It is also noteworthy that the patient responded well to high-dose daptomycin as defervescence occurred, only after having switched the antimicrobial chemotherapy from oxacillin to daptomycin.

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