Methicillin resistant staphylococcus aureus (MRSA) septicemia in pemphigus foliaceus - A coalition partners in resistant cases

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Abstract

Infection contributes to considerable morbidity and mortality in patients with autoimmune bullous disorders (AIBDs) such as pemphigus. Increased susceptibility to infection in this population may occur via 3 primary mechanisms- defective barrier function as a result of blistering and subsequent erosions, down regulation of immunity due to medications used in the treatment of AIBDs and finally, immune dysregulation associated with autoimmunity. In pemphigus, sepsis is the most common cause of death, with staphylococcus aureus being the most frequently implicated organism. We hereby report two cases of methicillin resistant staphylococcus aureus (MRSA) septicemia in pemphigus foliaceus (PF) patients rendering them resistant to conventional immunosuppressive therapies.

Key words

MRSA, pemphigus foliaceus.

Introduction

Infection contributes to considerable morbidity and mortality in patients with autoimmune bullous disorders (AIBDs) such as pemphigus. Increased susceptibility to infection in this population may occur via 3 primary mechanisms- defective barrier function as a result of blistering and subsequent erosions, down regulation of immunity due to medications used in the treatment of AIBDs and finally, immune dysregulation associated with autoimmunity. In pemphigus, sepsis is the most common cause of death, with staphylococcus aureus being the most frequently implicated organism. We hereby report two cases of methicillin resistant staphylococcus aureus (MRSA) septicemia in pemphigus foliaceus (PF) patients rendering them resistant to conventional immunosuppressive therapies.

Case reports

Case 1 A 65-year-old male presented with crusting and erosions over the body since 20 days (Figure 1a). Nikolsky’s Sign was positive. Patient was clinically suspected to be suffering from pemphigus foliaceus (PF); diagnosis was confirmed by direct immunofluorescence (DIF) and desmoglein (Dsg) ELISA (Dsg1-200 IU/ ml; Dsg3-negative). He was started on oral prednisolone (1mg/ kg/ day) and azathioprine (1 mg/ kg/ day) along with intravenous amoxicillin-clavulanic acid combination and amikacin; the response to treatment was suboptimal than expected. Patient’s condition worsened in the next few days. Blood culture showed growth of methicillin resistant staphylococcal aureus (MRSA) for which patient
Case 2 A 39-year-old lady presented with extensive crusting and oozing over scalp, back, chest, abdomen and upper limbs. A clinical diagnosis of PF was made which was further confirmed by DIF and indirect immunofluorescence and Dsg ELISA showing desmoglein (>200 IU). Patient was put on prednisolone (1mg/ kg/ day) along with azathioprine (50mg/ day) under the cover of intravenous antibiotic (amoxicillin-clavulanic acid combination and amikacin). Repeat blood culture after 7 days revealed growth of MRSA sensitive to linezolid and clindamycin. Patient was given antibiotics parenterally. She responded dramatically in the next 5 days.

PF and Staphylococcus aureus both target extracellular domain of desmoglein-1. The Extracellular domain of Dsg-1 consists of 4 regions (EC1-EC4). The cause of blistering in staphylococcal infection is the exfoliative toxin which targets extracellular domain of Dsg1 whereas in PF it is due to production of autoantibodies. We hypothesize that there was twin attack on Dsg 1 in our patients –one from autoantibodies and other from toxin produced by MRSA. Autoantibodies in PF target extracellular domains (EC1) regions while staphylococcal toxin cleaves Dsg1 through its action at the junction between EC3 and EC4 region of Dsg-
In such a scenario, patients might not respond optimally to standard immunosuppressive regimen if the underlying infection is not treated efficiently. It is possible that physician not being aware of the hidden staphylococcal infection might overload the patient with higher doses of steroids and/or steroid sparing agents when the expected response is not achieved after putting them on the specific therapy. It is therefore necessary to scrutinize for the concomitant infection (by repeating wound and blood culture) in PF patients who do not respond adequately to standard regimen. Moreover, overt signs of septicemia like high grade fever may not be seen in patients who are receiving high dose of steroids. The empirical antibiotic therapy in AIBDs covers gram positive bacteria including staphylococcus aureus under normal circumstances; however, the problems arises in cases of infections with MRSA which doesn’t respond to the empirical antibiotic therapy. Therefore, practitioners must have a high index of suspicion to evaluate for the underlying focus of infections in patients with pemphigus who fails to respond to, or worsen on, systemic immunosuppressive medications. Appropriate antibiotic therapies must be instituted in these patients before up scaling the dose of immunosuppressive therapy.

References