Atypical DRESS Syndrome in a Post-Partum Patient

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Received date: 24-11-2015; Accepted date: 2-11-2015; Published date: 5-12-2015

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Introduction
A 35-year-old female 14 days postpartum was evaluated in the hospital for a diffuse pruritic papular rash. Her peripartum course was complicated by preeclampsia and liver rupture necessitating emergent caesarean-section with exploratory laparotomy and partial heptectomy. Subsequently, she developed coagulase-negative staphylococcal bacteremia and was discharged with a 2-week vancomycin course. Four days after antibiotic completion, she developed diffuse erythematous, pruritic papules over her trunk and abdomen (Figure A) which progressed to the face, arms, back, and legs. She then developed systemic symptoms of fevers, nausea, and emesis prompting hospitalization. On presentation, she was restarted on vancomycin for concerns of recurrent infection and prednisone for relief of presumed urticaria.

On physical examination, she was febrile (38.7°C) with facial swelling, angular cheilitis, and axillary lymphadenopathy. She had partially blanchable pink papules over the face, arms, legs, and trunk. No mucous membrane involvement was noted.
Laboratory evaluation revealed a normal white blood cell count (5.7x10³/μL) with atypical lymphocytes (2%, 110 cells/mm³) on differential (Figure B). There was no peripheral eosinophilia, likely due to steroid use. A comprehensive metabolic panel demonstrated acute kidney injury (creatinine 1.4mg/dL, baseline of 0.8mg/dL). Liver function was normal. HIV antibody and PCR were negative.

Figure B. Atypical lymphocyte on peripheral blood smear

Discussion

Given the patient’s rash, systemic symptoms, atypical lymphocytosis, and timing of vancomycin administration, drug reaction with eosinophilia and systemic symptoms (DRESS) was suspected. DRESS syndrome is a potentially fatal drug hypersensitivity reaction characterized by acute rash, hematologic abnormalities, lymphadenopathy, and other organ involvement that occurs 2-8 weeks after drug exposure. Acute rash may take varying forms including erythematous macular lesions that first appear on the face, upper trunk, and upper extremities which may progress to vesicles, purpura, or exfoliative dermatitis. While a majority of cases demonstrate peripheral eosinophilia, atypical lymphocytosis is often noted. Hepatic and renal dysfunction are common.

The European Registry of Severe Cutaneous Adverse Reaction (REGIscar) study group proposed diagnostic criteria for DRESS which include three required criteria (acute rash, suspicion of drug-related reaction, and hospitalization) as well as at least 3 of 4 systemic features (fever, lymphadenopathy involving 2 sites, involvement of at least 1 internal organ, and hematologic abnormalities). Our patient had a REGIscar score of 4, indicating probable DRESS. As reactivation of human herpesviruses has been suggested as an etiology of DRESS syndrome, serologies to HHV6, HHV7, CMV, and EBV were sent and all negative.

While the most frequent causative agents of DRESS are anticonvulsants, other medications including sulfonamides, vancomycin, and allopurinol have been reported. Vancomycin, the likely causative agent, was discontinued and the patient was treated with oral prednisone with complete resolution of symptoms within one week. The patient and her first-degree relatives were educated about increased risk for DRESS occurrence with vancomycin and structurally related medications.

In summary, DRESS syndrome is potentially life-threatening with significant morbidity. Prompt diagnosis is vital along with identification and withdrawal of suspect medications. Clinicians should be aware of the varying clinical presentations of DRESS syndrome, including the likelihood of masking peripheral eosinophilia with steroid administration.

Acknowledgements:

Tara Shankar MD and Merritt L. Fajt MD had full access to all of the data in the study and take responsibility for the integrity of the data and the accuracy of the data analysis. Division of Pulmonary, Allergy and Critical Care Medicine, Department of Medicine, University of Pittsburgh Medical Center, Pittsburgh, Pennsylvania, USA.
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