THE DEADLY DRESS
Jason Vuong, BA; Gwendolyn Ho, MD; Edris Aman, MD; Gerald Diaz, MD; Faith Fitzgerald, MD
University of California, Davis Medical Center, Sacramento, CA

LEARNING OBJECTIVES

• Drug Reaction with Eosinophilia and Systemic Symptom (DRESS) is a rare, potentially life-threatening syndrome.
• Patients often present 2-6 weeks after the start of drug therapy with skin rash, liver involvement, fever, hypereosinophilia, and lymphadenopathy.
• Incidence of 1 in 1000-10,000 drug exposures.
• DRESS carries up to a 10% mortality rate.
• Early detection and treatment is critical.

CASE PRESENTATION

A 48-year-old man with a history of epilepsy presents with a two-week history of rash, fever and diffuse lymphadenopathy and severe facial swelling.

Four weeks prior to admission, his anticonvulsants had been switched from phenytoin to levetiracetam and carbamazepine.

Two weeks following the medication change, he developed a morbilliform rash that rapidly progressed.

On presentation, he was persistently febrile with diffuse tender lymphadenopathy and follicular erythematous papules on the face, trunk, and extremities with sparing of the palms and soles of the feet.

His labs were remarkable for leukocytosis of 11 with peripheral eosinophilia and elevated transaminases. A peripheral blood smear showed atypical, reactive lymphocytes. Infectious workup was unremarkable. A skin biopsy revealed folliculitis with eosinophils consistent with a drug reaction attributed to his new anticonvulsants.

He was started on oral and topical steroids and had immediate clinical improvement in his symptoms but his hospital course was prolonged by persistent fevers and elevating transaminases that improved with higher doses of systemic steroids; thereby allowing for discharge on HD#7 with a prednisone taper and topical steroids.

LABS

<table>
<thead>
<tr>
<th>Temp</th>
<th>WBC</th>
<th>Absolute Eosinophil Count</th>
<th>ALT &amp; AST</th>
</tr>
</thead>
<tbody>
<tr>
<td><img src="chart1.png" alt="Temp Chart" /></td>
<td><img src="chart2.png" alt="WBC Chart" /></td>
<td><img src="chart3.png" alt="Absolute Eosinophil Count Chart" /></td>
<td><img src="chart4.png" alt="ALT &amp; AST Chart" /></td>
</tr>
</tbody>
</table>

DISCUSSION

DRESS syndrome was initially described as anticonvulsant hypersensitivity syndrome because anticonvulsants accounted for one third of drugs causing DRESS. Altogether, 44 total drugs have been associated with DRESS.

This patient presented with a classic case of DRESS syndrome secondary to carbamazepine. DRESS syndrome commonly develops 2-6 weeks after exposure to the inciting drug with symptoms of fever, liver involvement, hypereosinophilia, lymphadenopathy, and rash. The rash begins as a morbilliform eruption, which later becomes edematous with follicular accentuation that may persist for weeks to months after drug withdrawal. Accompanying facial edema occurs in 39% of DRESS cases. The liver is the most common, and most severe site of visceral involvement. Because 10% of DRESS patients can develop a fulminant hepatitis that carries a high associated mortality rate, serial measurements of liver enzymes are necessary. The kidneys and lungs are the next most common sites of involvement.

The lack of a rapid diagnostic test makes having a high index of suspicion crucial for diagnosis. The RegiSCAR’s scoring system may aid in the recognition of DRESS syndrome in at-risk patients and can help prevent progression of the syndrome. This scoring system can be used to classify the probability that a patient has DRESS. In our patient, his RegiSCAR score was 7, which places him in the “definite” category of having DRESS and, fortunately, early therapy was initiated, preventing any significant complications.

REFERENCES


CONTACT

Jason Vuong, MS3
jason.vuong@ucdmc.ucdavis.edu
UC Davis School of Medicine