DRESS Syndrome-An Unusual Presentation with Pancytopenia after Salazopyrin Treatment

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Abstract

**Background:** Drug Reaction with Eosinophilia and Systemic Symptoms (DRESS) syndrome is a rare and potentially life-threatening adverse drug reaction. This syndrome is most frequently caused by various drugs such as allopurinol, Salazopyrin, antiepileptics (carbamazepine, lamotrigine, phenytoin, and phenobarbital), and antibiotics (sulfamethoxazole, dapson, minocycline, and vancomycin). Treatment usually includes discontinuation of the offending drug and initiation of high-dose steroids with a prolonged taper.

**Objective:** To describe an unusual case of DRESS syndrome with pancytopenia during the time of initial presentation, following a late appearance of rash and eosinophilia.

**Case Presentation:** A 44-year-old woman was hospitalized in our internal medicine ward one month after Salazopyrin therapy was initiated. A thorough investigation for malignant or viral etiology was performed and the lab results raised the suspicion of Hemophagocytic Lymphohistiocytosis (HLH). Subsequently, eosinophilia, atypical lymphocytes and a rash appeared 10 days after her initial presentation and DRESS syndrome was diagnosed.

**Conclusion:** DRESS syndrome is a unique, potentially life-threatening, entity that can present in the form of fever and pancytopenia with delayed appearance of rash, eosinophilia and atypical lymphocytes. The recent use of drugs described as related to DRESS should raise the preliminary suspicion. In order to avoid complications, a high index of suspicion is warranted.

**Keywords:** DRESS syndrome, Salazopyrin, Eosinophilia, Pancytopenia, HLH

Introduction

Drug Reaction with Eosinophilia and Systemic Symptoms (DRESS) syndrome is a rare but potentially life-threatening adverse reaction that can result from exposure to a variety of medications. It most often develops after the use of sulfinilamide, allopurinol, phenytoin or other anticonvulsants. The syndrome is characterized by a late onset, usually occurring 2-6 weeks after exposure of the culprit drug, and a prolonged course. The clinical presentation varies greatly and includes a combination of fever, skin eruptions, lymphadenopathy, hematologic abnormalities (most often eosinophilia and atypical lymphocytes) and internal organ involvement [1]. As this presentation can mimic other conditions, mostly infection, diagnosis requires a high index of suspicion. We report a case of a 42-year-old woman who presented to our ward 4 weeks after initiation of Salazopyrin therapy, with fever, erythematous rash, pancytopenia, lymphadenopathy and hepatitis.

**Case Presentation**

A 44-year-old woman presented to the emergency department at our medical center after 4 days of weakness, fever (39.5°C) and lymphadenopathy. No additional complaints were noted. A few months before her admission, she underwent medical investigation for sudden arthralgia. Salazopyrin therapy was initiated a month prior to the current hospitalization due to suspected spondyloarthropathy. Prior to hospitalization, she was treated with amoxicillin-clavulanic acid, dindamycin and corticosteroids by her family physician, with no improvement. On physical examination, the
DRESS syndrome is a potentially life-threatening syndrome that includes cutaneous eruptions, fever, hyper-eosinophilia and internal organ involvement (lungs, kidneys, liver) [2]. The estimated incidence of this syndrome ranges from 1 in 1,000 to 1 in 10,000 drug exposures [3]. It is considered a dermatological emergency with a mortality rate of approximately 10% [4]. The initial presentation of DRESS syndrome can often be misleading. The variability of initial features led to the development of the European Registry of Severe Cutaneous Adverse Reaction Criteria (RegiSCAR) scoring system, which is commonly used to aid diagnosis [5].

The patient described in this case first presented with high fever and lymphadenopathy four weeks after starting on Salazopyrin. Blood tests demonstrated pancytopenia and elevated liver enzymes, without hyper-eosinophilia or atypical lymphocytes. Only a week after initial presentation and 4 weeks after Salazopyrin was discontinued, a typical rash with marked eosinophilia and high, atypical lymphocyte count erupted. Despite this unusual presentation, a definitive diagnosis of DRESS syndrome was made according to the RegiSCAR scoring system: fever, lymphadenopathy, eosinophilia, elevated liver enzymes (as a marker for hepatitis and liver involvement), a rash that involves more than 50% of the body surface and facial edema. Surprisingly, the features presented by this patient also answer the 2004 revised diagnostic criteria for HLH-fever, pancytopenia, elevated sIL-2R levels and reduced natural NK cell activity [6]. The overlap of DRESS and HLH were described in a few case reports in the literature [7-11]. It is still unclear whether HLH is a complication of DRESS or a situation that “mimics” this unique syndrome [12]. Regardless, both reflect T cell hyper-activity and are treated with trigger removal and administration of systemic steroids [13].

The mainstay treatment of DRESS syndrome includes withdrawal of the suspected culprit drug and the use of systemic corticosteroids. High index of suspicion is warranted, since if administered in the acute stage of the disease, corticosteroids may improve the patient’s long-term outcome [14]. We described the presentation and disease evolution of a 44-year-old patient treated with Salazopyrin, which is among the most common causes of DRESS syndrome. The diagnosis was delayed due to atypical presentation in the form of pancytopenia, which was probably an adverse reaction to Salazopyrin, late skin eruption and belated appearance of peripheral blood eosinophilia.

Conclusion

DRESS syndrome is a rare, potentially life-threatening, adverse reaction to various medications. Symptoms include skin rash, fever, lymphadenopathy, liver involvement and hyper-eosinophilia. High index of suspicion is warranted, since systemic therapy is often indicated in-order to ensure the affected patient’s safety.
References


