

Case Report

Dress syndrome – A case report with myriad systemic manifestations

¹Dr. Anagha. R. Auradkar , ²Dr. Suresh G*

¹Junior Resident, ²Professor

Department of General Medicine, K.S.Hegde Medical Academy,

NITTE University, Mangalore, India.

Corresponding author*

ABSTRACT

Drug rash with eosinophilia and systemic symptoms syndrome is a severe idiosyncratic drug reaction with a long latency period. It has been described using many terms; however, drug rash with eosinophilia and systemic symptoms syndrome appears to be the most appropriate. This syndrome causes a diverse array of clinical symptoms, anywhere from 2 to 8 weeks after initiating the offending drug. We report a case of generalized macular rash, pruritis, pleural effusion and acute hepatitis secondary to amoxicillin and sulphasalazine (sulpha drugs)

Key words : Drug reaction ; Eosinophilia ; systemic symptoms

INTRODUCTION

Drug reaction with eosinophilia and systemic symptoms (DRESS) syndrome, also known as drug-induced hypersensitivity syndrome (DIHS), is an under-recognized and potentially life-threatening hypersensitivity reaction associated with a variety of medications, many being anti-epileptics. Patients with DRESS syndrome typically present with rash, swelling, fever, and systemic manifestations such as a severe transaminitis. In most cases, a patients face, trunk, and upper extremities are affected by a rash which is at first morbilliform then gradually transitions to maculopapular, and finally can progress to edema of the face, particularly in the periorbital region. Although rash and eosinophilia are commonly seen in hypersensitivity reactions, the defining characteristic of DRESS syndrome is organ dysfunction, most commonly of the liver, kidneys, heart, or lungs.

CASE REPORT

A 25year old female patient presented to us with fever associated with generalised pruritic macular rash of 12 days duration. Prior to presenting to our hospital: 2 months ago she was diagnosed with Psoriatic arthritis and was initiated on Sulphasalazine and HCQS (hydroxychloroquine) which she took for one month and sulphasalazine for two weeks. 2 weeks prior she had one day history of fever for which she had been treated with oral amoxicillin and the second day, she developed anaphylactic shock with sepsis and was referred to our centre for further management. At presentation, her total counts were elevated with eosinophilia (TC= 69,000cell/cu.mm, eosinophils – 15%, AEC – 6336 cell/ cu.mm) with acute hepatitis (SGOT – 2475, SGPT – 941, total bilirubin 6.98mg/dl). She was treated with ionotropic support and initiated on Inj. Meropenem. Peripheral smear showed no evidence of malignancy. Patient recovered gradually with parenteral antibiotics and supportive care. Bone marrow showed hypercellular marrow with increase in eosinophilic precursors. USG abdomen was suggestive of mild hepatomegaly and chest x-ray and ECG were normal. A skin biopsy for her macular rashes was done which showed lymphocytic small vessel vasculitis and she was initiated on oral steroids. The constellation of eosinophilia, generalized macular rashes on the face trunk and extremities associated with elevated transaminases, favoured the diagnosis of DRESS (Drug reaction, eosinophilia and

systemic symptoms)During the course of stay she developed right sided pleural effusion, for which she was underwent ICD insertion and pleurodesis following the drainage of pleural fluid. Patient also developed transient conduction abnormality (ECG showed AV dissociation). 2 weeks following these symptoms, she developed right ear external otitis media and was treated with oral ciprofloxacin. There was no exacerbation following initiation of this drug and she was discharged with oral steroids (Sulphasalazine was discontinued). 3 weeks later, at follow up the liver enzymes had normalized, Chest x-ray showed no evidence of pleural effusion and ECG showed sinus rhythm with no conduction abnormalities. There has been no recurrence reported till date (8 months since discharge)

DISCUSSION

Drug reaction with eosinophilia and systemic symptoms syndrome is a potentially life-threatening hypersensitivity reaction with rash, fever, and internal organ involvement, often hepatitis, occurring most commonly two to eight weeks after initiation of a medication. It was first described in 1936 during treatment with anticonvulsant drugs. Later on, the association with other drugs was established and the name 'DRESS syndrome' was suggested to describe this entity. The syndrome is characterised by rash, fever, lymphadenopathy and internal organ involvement (single or multiple). The pathogenesis is not fully understood. It has been suggested that certain drugs may cause a hypersensitivity reaction as a result of abnormalities in the production and detoxification of its active metabolites in patients with genetic or acquired variations in drug metabolism pathways. Its incidence ranges between 1 in 1000 and 1 in 10 000 exposures. The aromatic anticonvulsants(phenytoin, phenobarbital, carbamazepine) and sulphonamides are the most common drugs described in this syndrome, but presentation involving multisystem is rare. The most commonly involved organ is the liver. Diagnostic criteria for DRESS syndrome, published in 1996 by Bocquet *et al* , include the simultaneous presence of three conditions:

- ▶ Drug-induced skin eruption
- ▶ Eosinophilia $\geq 1500/\text{mm}^3$ and

At least one of the following systemic abnormalities:

- ▶ Lymphadenopathy
- ▶ Hepatitis (transaminases >2 ULN)
- ▶ Interstitial nephropathy
- ▶ Interstitial lung disease
- ▶ Myocardial involvement.

The skin biopsy may help to confirm the diagnosis but is usually not specific. It shows a lymphocytic infiltrate of the papillary dermis, which may contain eosinophils and is generally denser than in other drug reactions. The most common differential diagnoses include Stevens–Johnson syndrome (SJS), toxic epidermal necrolysis (TEN), hypereosinophili syndrome and Kawasaki disease. So far, prompt withdrawal of the offending drug is the only undisputed way to treat drug hypersensitivity reactions. Supportive therapy includes antipyretics and the use of topical steroids to improve symptoms. Systemic corticosteroids can reduce symptoms of delayed hypersensitivity reactions.

CONCLUSION

Given the significant mortality attributed to drug reaction with eosinophilia and systemic symptoms syndrome, medical personnel should be aware of the potential for this severe hypersensitivity reaction and should ensure close follow-up and offer anticipatory guidance when beginning any new medication, particularly anti-epileptic therapy. Early recognition of drug reaction with eosinophilia and systemic symptoms syndrome and initiation of appropriate therapy are imperative in limiting morbidity.

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