Carbamazepine Induced DRESS Syndrome

Efnan Çalık[1], Betül Çavuşoğlu Türker[1], Süleyman Ahbab[1], Hayriye Esra Ataoğlu[1]

ABSTRACT

Introduction: The DRESS (Drug Reaction with Eosinophlia and Systemic Symptoms) Syndrome, is a rare encountered disease with delayed type hypersensitivity reaction, induced by drugs which accompanied by fever, skin rash, lymphadenopathy and internal organ involvement.

Case report: It is presented a 29-year-old female patient with DRESS syndrome who admitted to the hospital such complaints as fever, extensive maculopapular rash, submandibular lymphadenopathy, hypertransaminasemia, eosinophilia due to carbamazepine treatment.

Conclusion: The most common cause of The DRESS Syndrome is carbamazapine. The drug must be stopped and prednisolone should be given. Early diagnosis and early discontinuation of the drug significantly reduce mortality.

Keywords: Carbamazepine, Drug Reaction, Eosinophilia, DRESS Syndrome

INTRODUCTION

DRESS (Drug Reaction with Eosiophilia and Systemic Symptoms) syndrome severe acute drug induced hypersensitivity reaction. It is life-threatening disease and mortality rate is about 10% which is mostly due to liver disfunction. The estimated incidence of this syndrome ranges from 1 in 1000 to 1 in 10,000 drug exposures[1]. It is caused by drugs such as anticonvulsants (carbamazepine, phenobarbital, phenytoin), sulfonamids, allopurinol, antimicrobial agents, lamotrigine, sulfasalazine. Clinical features begin 3-8 weeks after commencement of drug. DRESS syndrome is characterised by diffuse maculopapular rash, facial edema, lymphadenopathy, fever, hematologic abnormalities (eosinophilia), involvement of internal organs such as liver, heart, lungs. The pathogenesis of DRESS syndrome has been still unclear but probably involves different mechanisms including detoxification defects, genetic predisposition to immune reactions, and the reactivation of human herpes, Epstein–Barr virus, cytomegalovirus, HHV-6 and -7. It should be thougt neoplastic diseases (lymphoma, leukemia, hypereosinophilic syndrome, paraneoplastic), autoimmune diseases or connective tissue conditions (adult-onset Still's disease, lupus erythematosus, vasculitis) in differential diagnosis. Stevens Johnson, toxic epidermal necrolysis, hypereosinophilic syndrome should be thought as differential diagnosis too. We describe a case of a 29-year old woman admitted to our hospital with acute diffus rash, fever, submandibular lymphadenopathy six weeks after starting carbamazepine therapy.

CASE REPORT

A 29-year old woman was being followed in the neurology outpatient clinic for trigeminal neuralgia. For this reason, carbamazepine was prescribed. After six weeks of carbamazepine treatment she was admitted to our clinic with acute diffuse skin rash, face edema, mainly periorbital, fever, submandibular and servical lymphadenopathy. On examination the patient was conscious and oriented, body temperature was 37.8°C. Rash was erythematous and started from the chest and progressed to involve other body parts as shown in figure 1 and 2. Patients's informed consent and permission to use the photograph were provided. Cervical and submandibular lymphadenopathy was also present. Laboratory tests showed white blood cell count (8400cell/µL) with eosinophlia 11%, with no atypical lymphocytes, liver parameters values: aspartate transaminase (AST) 322 IU/L; alanine transaminase (ALT) 331 IU/L; alkaline phosphatase (AP) 539 IU/L; gamma-glutamyl transferase (GGT) 734 IU/L; lactate dehydrogenase (LDH) 240 U/L; total bilirubin 4,3 mg/dL, INR 1,74 serum creatinine level of 0,75 mg/dL; serum urea level of 35 mg/dL; C-reactive protein 19 mg/dL. Serologic tests for acute infection by human immunodeficiency virus, hepatotropic virus – hepatitis A, B, C, E, Epstein–Barr virus, Citomegalovirus, Toxoplasma,VDRL were negative. Antinucleer antibody (ANA), antimitochondrial antibody (AMA), anti Liver kidney microsome (anti-LKM), Anti Smooth Muscle Antibody (ASMA) were negative. Dermatology was consulted and considered the skin lesions could be DRESS syndrome induced by carbamazepine and a biopsy of one of the skin lesions was performed. On the thirth day of hospitalization INR value raised 3. Consulted with hospital's gastroenterologist and prednisolone, IVIG therapy were initiated. The diagnosis of DRESS syndrome was made (using RegiSCAR criteria, Calculated RegiSCAR score was 4, (acute skin rash, fever, involvement of liver, and eosinophils above the laboratory limits). prednisolone was increased to 1 mg/kg/day. After starting prednisolon and IVIG therapy liver enzymes reduced gradually. Skin lesions improved progressively. The patient was discharged home with the reduce corticosteroid therapy gradually.

[1]Health Sciences University, Haseki Training and Research Hospital, Internal Medicine Clinic, Turkey

Corresponding author: Betül Çavuşoğlu Türker, Adress: Health Sciences University, Haseki Training and Research Hospital, Internal Medicine Clinic, Fatih – İstanbul, Turkey

How to cite this article: Efnan Çalık, Betül Çavuşoğlu Türker, Süleyman Ahbab, Hayriye Esra Ataoğlu. Carbamazepine induced DRESS syndrome. International Journal of Contemporary Medical Research 2017;4(12):19-20.

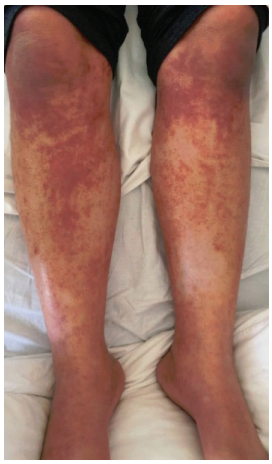

Figure-1: Erythematous skin lesions on lower extremities

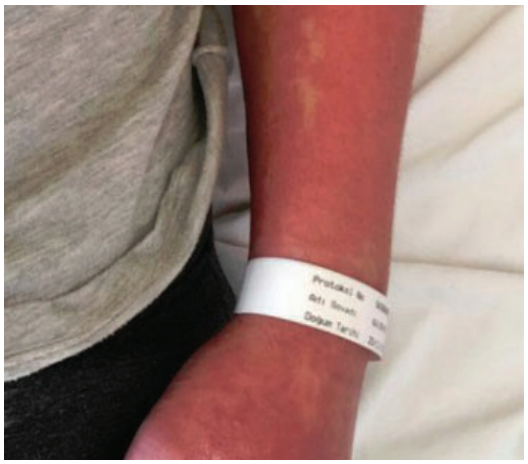

Figure-2: Erythematous skin lesions on arm

DISCUSSION

DRESS (Drug Reaction with Eosiophilia and Systemic Symptoms) syndrome fatal acute hypersensitivity reaction induced by drugs. It is caused by drugs such as anticonvulsants (carbamazepine, phenobarbital, phenytoin), sulfonamids, allopurinol, antimicrobial agents, lamotrigine, sulfasalazine. The most commonly reported drug is carbamazepine[2]. Diagnosis of DRESS syndrome is based on clinical features. There is no gold standart test for diagnosis. Laboratory tests can help to differentiate DRESS syndrome from other illnesses. The diagnosis is based on scoring system. The most common scoring system is RegiSCAR (the European Registry of Severe Cutaneous Adverse Reactions). RegiSCAR criterias are acute rash, fever above 38°C, lymphadenopathy at two sites, the involvement of at least one internal organ, and hematologic abnormalities[3]. In the present report, our patient developed all 4 criteria of the DRESS syndrome.

Clinical features are fever, skin rash, lymphadenopathy, involvement of internal organs.The liver most commonly affected organ[4]. Liver failure is main cause of death.It can vary from hepatitis to fulminan hepatic failure. We found a significant elevation in liver enzymes. DRESS syndrome can also involve other internal organs such as the kidney, lung, heart. There was not any other organ dysfunction in our case. Fever, eosinophilia, cervical and submandibular lymphadenopathy was detected. The most common laboratory finding among hematologic abnormalities is eosinophilia. High fever and lymphadenopathy are common symptoms of DRESS syndrome[5].

CONCLUSION

In conclusion, DRESS syndrome is a life-threatening delayed-type drug hypersensitivity reaction with 10% mortality rate. DRESS should be suspected in a patient who has skin rash, high fever, facial edema, lymphadenopathy and involvement of internal organ. Treatment should be started immediately. The drug must be stopped and prednisolone should be given. Early diagnosis and early discontinuation of the drug significantly reduce mortality.

REFERENCES

1. Cacoub P, MusetteP, Descamps V, et al: the DRESS syndrome: A literature review. Am J Med 2011;124:588-97
2. Kardaun SH, Sekula P, Valeyrie-Allanore L, Liss Y, Chu CY, Creamer D, et al. RegiSCAR study group. Drug reaction with eosinophilia and systemic symptoms (DRESS): An original multisystem adverse drug reaction. Results from the prospective RegiSCAR study. Br J Dermatol 2013;169:1071-80.
3. Mockenhaupt M. Epidemiology of cutaneous adverse drug reactions. Chem Immunol Allergy. 2012;97:1-17
4. Criado PR, Criado RFJ, Avancini JM, Santi CG: Drug reaction with eosinophilia and systemic symptoms (DRESS)/ drug-induced hypersensitivity syndrome (DIHS): a review of current concepts. An Bras Dermatol 2012; 87: 435-49.
5. Walsh S, Diaz-Cano S, Higgins E, et al: Drug reaction with eosinophilia and systemic symptoms (DRESS): Is cutaneous phenotype a prognostic marker for outcome ? A review of clinicopathological features of 27 cases. Br J. Dermatol 2012 Oct3.

Source of Support: Nil; **Conflict of Interest:** None

Submitted: 09-12-2017; **Accepted:** 07-01-2018; **Published:** 16-01-2018