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Case Report

Dapsone induced DRESS syndrome: a rare fatal complication

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ABSTRACT

DRESS syndrome is a serious life threatening condition characterized by skin eruption, haematological abnormalities and multi organ involvement that can be fatal if unrecognized especially in patients with liver failure. Diagnosis may be difficult because it is rarely seen in children and it can mimic many different conditions. Author report a case of 12 year old female presented to this emergency department with moderate grade fever, skin rash and jaundice following dapsone ingestion. She was evaluated and was diagnosed as DRESS syndrome and successfully treated with steroids.

Keywords: Dapsone, Drug rash eosinophilia and systemic symptoms, Hypersensitivity

INTRODUCTION

Dapsone is the parent compound of sulfone drugs, used in the treatment of immumological, hypersensitivity and various skin disorders. It has anti-inflammatory and antibacterial activity.[1] However, the use of dapsone is associated with a myriad of idiosyncratic adverse effects like agranulocytosis, peripheral neutropathy, dapsone hypersensitivity or Drug rash, eosinophilia and systemic symptoms (DRESS) syndrome, primary hepatocellular and cholestatic hepatitis, permanenent retinal damage and haemolytic anemia. DRESS syndrome, represents a rare but acute and potentially fatal multisystem drug reaction.It is characterized by the clinical tetrad of fever, maculopapular rash, lymphadenopathy and internal organ involvement. Author report a case of 12 year old girl who presented with fever, jaundice and rash two weeks following dapsone ingestion.

CASE REPORT

A 12 year old female child was brought to the emergency with history ofmoderate grade, intermittent fever for 2 weeks. Yellowish discolouration of eyes was noted on second day of fever associated with high coloured urine. After 7 days of fever, parents noticed generalised rash which started on face (Figure 2) and progressed all over the body. Initially papular rash was present which progressed to a confluent erythematous rash. Peeling of skin was noted since 2 days prior to admission. Child was diagnosed as leprosy in view of history of a single hypopigmented lesion on face one month prior to this illness and was started on dapsone (100mg once daily). Dapsone was stopped by the parents after the onset of febrile illness.

On general examination mild pallor andicterus was present. Cervical lymphadenopathy of 1.5cm size, soft, freely mobile was present associated with bilateral eye congestion around limbus. No lesions or any ulcers were noted in oral cavity. Diffuse erythroderma was present over trunk and abdomen (Figure 1). Macular erythematous lesions were present on bilateral lower limbs. Peeling of skin over face was present. Systemic examination was unremarkable.

Complete blood count showed haemoglobin- 12.1gm%, Total count -9,700cells/mm^3 (45% Neutrophils, 50% Lymphocytes, 3% Eosinophils, 2% Monocytes), platelet count -2.59lac/mm^3. C-Reactive protein (CRP) was

normal. Liver function tests suggestive of elevated transaminases (SGOT-217 U/L, SGPT-259 U/l), ALP-1211 U/L, serum albumin-3.4 mg/dl with raise in total bilirubin level-3.3 mg/dl (direct bilirubin-2.4 mg/dl, indirect bilirubin-0.9mg/dl). ESR was 30mm/hr. Renal function tests, procalcitonin, coagulation profile were normal. Blood culture was sterile. Serology for HAV, HEV, HCV antibody were negative.

ANA profile was negative. A diagnosis of dapsone induced DRESS syndrome was made based on the clinical and laboratory parameters. Child was started on prednisolone at 35 mg/day (1 mg/kg/day) .Supportive care was given (Liquid paraffin lotion, hydroxizine was added for pruritus). Her fever spikes settled down in 3 days; rashes started to fade after a week. She was discharged on oral prednisolone for 2 weeks. On follow up child was doing well, skin lesions completely resolved with normalization of liver function tests (Total bilirubin -0.7mg/dl, SGOT -33U/L, SGPT - 4O U/L).

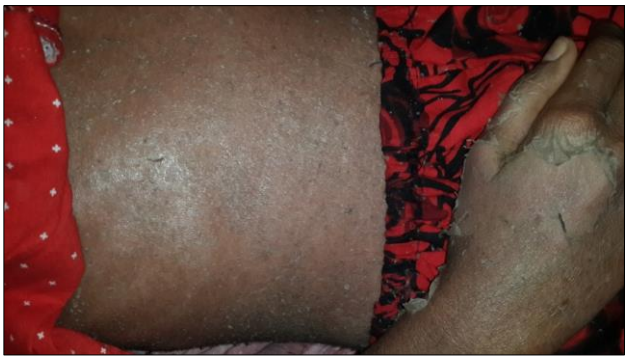

Figure 1: Diffuse erythroderma with exfoliation of skin present over abdomen and dorsum of hand.

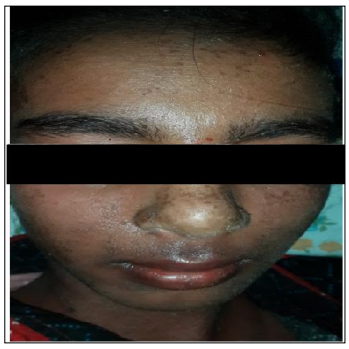

Figure 2: Diffuse erythroderma with exfoliation involving the face.

DISCUSSION

Drug rash with eosinophilia and systemic symptoms, is also called drug hypersensitivity syndrome or anticonvulsant hypersensitivity syndrome. It is caused by a T-cell response specific to the drug.[2] Reactivation of herpes virus, especially human herpes virus 6 also contributes to this syndrome via an unknown pathogenic mechanism. Most common triggers include anticonvulsant drugs like carbamazepine followed by sulphonamides.[3] Genetic predisposition with particular HLA allele types such as HLA-A *3101 has also been implicated with specific ethnic groups. Incidence ranges from 1 in 1000 exposure to 1 in 10,000 exposures.[4] It is more common in adult patients than children.[5]

It is classically seen 2-6 weeks after initial exposure.[6] They often manifest as tetrad of fever, rash, hepatitis andlymphadenopathy.[7] The skin rash initially starts on head, upper trunk and arms. A diffuse exanthem of pruritic, morbilliform papules is most common though any morphology may be present. Exofoliationearly in the course, as seen in toxic epidermal necrolysis, isuncommon. Prominentperiocular or facial edema, cervicallymphadenopathy, pharyngitis and malaise accompany this dramatic cutaneouseruption. Eosinophiliaand atypical lymphocytosis are common but not always present. Hepatitis may range from mild elevation of transaminases value to frank hepatic failure .Other complications include interstitial nephritis, pneumonitis, myocarditis, shock andencephalitis.[8]

Late onset thyroiditis and hypothyroidism may occur months later as a result of antimicrosomal antibodies directed against thyroid peroxidases involved in drug metabolism.[9,10] Because of its highly variable clinical presentation, other clinical conditions such as acute viral infections, hepatitis, sepsis, autoimmune diseases and hematological disorders should be considered in differential diagnosis of DRESS syndrome.

In this patient all typical signs and symptoms of this condition and biochemical abnormalities developed after the initiation of dapsone therapy. The Regiscar scoring system was used to achieve a definite diagnosis of DRESS syndrome. Withdrawal of the offending drug is the primary therapeutic intervention. Lymphocyte transformation tests and patch testing are helpful for identifying the offending drug when multiple suspect agents are present, but drug discontinuation should not be delayed while awaiting results.

Symptomatic treatment of pruritis and pain can be accomplished with emollients and mild to high potency topical corticosteroids. This may be sufficient to achieve the resolution of clinical and laboratory abnormalities in many children. In more aggressive cases corticosteroids or other immunosuppressive drugs should be considered to achieve the best outcome.

A high index of suspicion is needed for diagnosis and managing DRESS. Pediatrician should be well aware of this condition and be able to differentiate with close mimicres of the disease.

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