Dapsone Hypersensitivity Syndrome

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ABSTRACT

Dapsone hypersensitivity syndrome is a relatively uncommon disease observed after dapsone therapy. It is potentially serious disorder and high index of suspicion is necessary for diagnosis. Treatment of DHS is discontinuation of dapsone therapy and administration of steroids.

INTRODUCTION :

Dapsone hypersensitivity syndrome (DHS) is hypersensitivity reaction, manifesting with triad of fever, rash and organ involvement[1]. It is also called as'five week dermatitis' as it commonly occurs at five weeks after dapsone therapy. It may progress to dermatological emergency and rarely cause death.

CASE :

16 yrs old male patient was diagnosed to be a case of multibacillary Hansens' disease 34 days prior to admission and was prescribed multi drug therapy including dapsone. Patient presented to our center with history of fever with maculopapular rash over trunk which gradually spread over limbs since 20 days. Patient also had history of jaundice. On admission Patient was febrile (104 ^{0}F) , had tachycardia (110/min) , blood pressure was 110/70 mmHg, Icterus and pallor was present. Patient had cervical and inguinal lymphadenopathy. Maculopapular rash was present all over body. Tender hepatomegaly of 7 cm was present.DHS was suspected in view of history of Dapsone therapy and typical triade i.e. fever, jaundice and presence of rash .

Investigation revealed, anemia (Hb 7.7gm%), thrombocytopenia (platelet count 43000/mmcu), leucocytosis (TLC 20200/mmcu), lymphocytosis (53%) was also reported. Liver enzymes were also raised (SGOT 454IU, SGPT 427IU, ALP 219IU), total bilirubin was 6.24mg/dl, Kidney function test revealed raised creatinine (2.5mg/dl), Blood urea was also high (82.5mg/dl).

Patient was started on systemic steroids, inj. dexamethasone 4mg tid. Fever subsided on day 3 and repeat total leucocyte count, lymphocyte count and platelets were in the normal range. Maculopapular rash subsided on day 10 and exfoliation developed. Patient was discharged on oral steroids and was asked to follow up for tapering steroids and for treatment of Hansens disease (excluding Dapsone) .

DISCUSSION :

DHS is rare syndrome 1st described by All day, Lowe,Barnes[2,3], as hypersensitivity vasculitis syndrome[4]. It has a incidence of 0.5 to 3%[4]. Clinical manifestations include fever, malaise, exfoliative dermatitis, hepatitis, hemolytic anemia, lymphadenopathy, a typical lymphocytosis, multiorgan involvement[5]. Cutaneous lesion are always present[3].Fever is common manifestation in DHS and can be observed before the rash appears[6]. DHS can begin after prolonged exposure to Dapsone and is reported even after 6 months of Dapsone therapy[7]. However it is classically reported during the first 3 to 8 weeks of daily therapy[5].

Diagnosis is based on triad of clinical manifestations including rash, fever and systemic involvement in patients who are on Dapsone and is supported by investigation i.e. raised liver enzymes, ESR and increased serum bilirubin.

Our case had a classical presentation, fever and rash

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started 2 wks after initiation of dapsone. Icterus, lymphadenopathy, generalized rash and exfoliative dermatitis was noted. Investigations revealed raised liver enzymes, raised ESR, lymphocytosis and anemia. Response to steroid also supports the diagnosis of DHS.

Though exact mechanism not known, it is thought that defect in metabolism of Dapsone is the cause for DHS. Dapsone is normally metabolized by two primary path was viz- N- acetylation and N-hydroxylation. In cases of DHS, there is defective detoxification of metabolite of Dapsone. Formation of toxic intermediate metabolites such as nitrosoamines and other compound are responsible for hemolysis and hepatotoxicity[7].

Management includes immediate discountinuation of drug. Administration of systemic corticosteroids is reported to be the mainstay of therapy in severe cases. Steroids should be tapered of over a month. Dapsone should not be included when antileprosy regimen is restarted[7].

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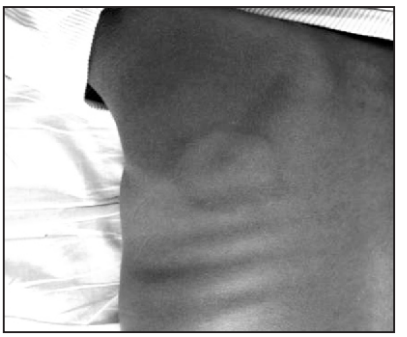
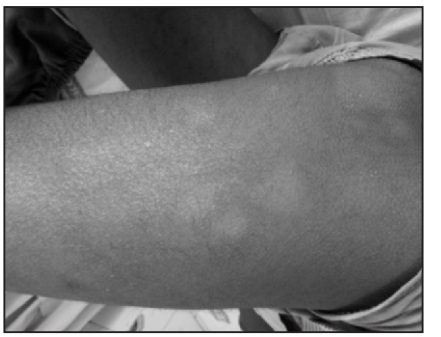
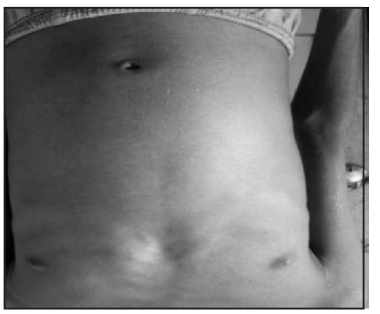


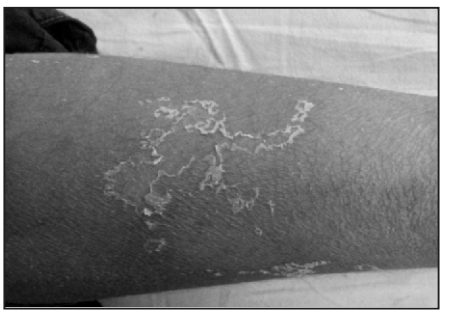
Hypopigmented patch over back



Multiple hypopigmented patches overthigh



Maculopapular rash over trunk



Exfoliative dermatitis