

Case Report

SULFONE SYNDROME – A CASE REPORT

A.M. Zende¹, U.Y. Bhoi², *R.R. Bhosale¹, N.K. Patankar¹ and S.N. Pradhan²

¹Department of Pharmacology, R.C.S.M. Govt. Medical College, Kolhapur, Maharashtra, India

²Department of Skin and V.D., Govt. Medical College, Miraj, Dist.Sangli, Maharashtra, India

*Author for Correspondence

ABSTRACT

Dapsone, a sulfone is an essential component of WHO recommended schedule for multidrug therapy of both paucibacillary and multibacillary leprosy. Dapsone is found to produce a specific type of hypersensitivity reaction in susceptible individuals named as “dapsone or sulfone syndrome”. Here is a case report of an 11 year old child with dapsone syndrome.

Key Words: Adverse Drug Reaction, Dapsone, Sulfone Syndrome

INTRODUCTION

Dapsone is a sulfone closely related with sulphonamides and has similar mechanism of action. It is leprostatic drug commonly used in treatment of both multibacillary (MB) and paucibacillary (PB) leprosy (Goodman and Gilman, 2001). Dapsone was introduced for the treatment of leprosy patients by Robert Chochrane in 1947 in Chingelput, South India (Jopling, 1998). Though variety of adverse drug reactions are associated with dapsone, the “dapsone syndrome” is distinct hypersensitivity reaction. The earliest report of hypersensitivity reaction to it was published in 1949 (Richardus, 1989).

CASES

An 11 year old child was diagnosed as a case of tuberculoid leprosy & was started with Tab.Dapsone 50 mg daily & Cap. Rifampicin 300 mg monthly single dose as per WHO schedule of multidrug therapy (MDT) for PB leprosy. He was investigated for G-6-PD deficiency.



Figure 1: Scaling and exfoliation of the skin

Case Report

After 4 weeks of the above therapy boy developed high grade fever with eruption of red coloured itchy rash all over the body. This was associated with vomiting, yellowish discolouration of sclera and swelling all over body. Patient was admitted in private hospital and received treatment in the form of i.v. fluids, antibiotics and liver supportive drugs. The drugs for PB leprosy were still continued. Ten days later when there was no relief, the patient was referred to nearby medical college hospital where on examination he was found to have generalized erythematous, maculopapular eruptions associated with lymphadenopathy, scaling & exfoliation of the skin (Figure 1). Patient also had dark yellowish discolouration of sclera & urine. His blood examination revealed Hb-10.1 gm%, WBC – 17800/cumm, Neutrophils – 59%, Lymphocytes – 31%, Eosinophils – 2%, Monocytes – 8%, Platelets – 83000/cumm. Total bilirubin was 9 mg/dl (Normal 1.0 mg/dl), SGPT 335 U/L (Normal 7-56 U/L), Serum Alkaline phosphatase 1368 U/L (Normal value for <15 year : <364 U/L). Abdominal ultra sonography showed hepatomegaly & splenomegaly. The patient was diagnosed as a case of sulfone syndrome and advised to stop dapsone. Patient received treatment initially with parenteral medications in the form of i.v. fluids, multivitamins, antibiotics, vitamin K, steroids and oral medications like syrup hepamerz 5ml 12 hourly, tab levocetizine. After 3 days parenteral steroids were stopped and patient was shifted on oral steroid therapy. Patient responded well to above therapy and there was dramatic improvement with reduction in itching, scaling, erythema and swelling of lymph nodes. The eye colour became normal and there was significant reduction in total bilirubin levels (4mg/dl) within 3 days which later became normal.

DISCUSSION

Dapsone is usually a well tolerated drug. However, it may produce non-hemolytic anemia and methaemoglobinaemia in person having G-6PD deficiency. Some other side effects include nausea, loss of appetite, pruritus, drug fever, reversible neuropathy and hepatotoxicity. Besides all these side effects, dapsone is also known to produce a symptom complex after 4-6 weeks of its ingestion. This consists of maculopapular rash, exfoliation, fever, lymphadenopathy, jaundice and heptasplenomegaly (Goodman & Gilman, 2001). These features were first described by Lowe and later Allday named this as “dapsone syndrome”. (Lowe, 1950; Allday, 1949) Dapsone syndrome is not a rare adverse drug reaction associated with use of dapsone as it was previously believed. Though the pathogenesis of dapsone syndrome is yet unknown, just withdrawal of drug had produced complete recovery in mild cases and other cases with additional supportive medications.

The present case of 11 year old boy who was diagnosed as a case of PB leprosy and treated with MDT as per schedule. But 4 weeks later the boy presented with all symptoms and signs of sulfone syndrome which started regressing after stoppage of dapsone. In present case as soon as the drug use was stopped, the boy started recovering and supportive medications hastened the recovery.

In geographical areas with high prevalence rates of leprosy, physicians should be aware to this severe and probably not so rare, hypersensitivity reaction to dapsone.

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