INTRODUCTION

Steven Johnson Syndrome (SJS) is a rare and potentially fatal immune complex hypersensitivity reaction that often presents as life threatening medical emergency. SJS and its more severe variant Toxic Epidermal Necrolysis (TEN) are caused by medications, although viral infections and malignancies have been reported.1

CASE REPORT

A 25 year old female patient presented with fever, generalized weakness, diffuse erythematous maculopapular rash all over the body with itching. There were erosions in oral and genital cavity. There was swelling and reddening of eyes with erosions on upper and lower eyelid and haemorrhagic discharge from both eyes. All these symptoms started 2 weeks after commencing Category II anti TB regimen consisting of rifampicin, pyrazinamide, isoniazid and ethambutol (HRZE) in fixed dose combination and intramuscular injections of streptomycin. Fifteen days after the commencement of Cat II anti TB treatment she developed diffuse erythematous rash on face, trunk and both extremities which turned into blisters. There were ulcers on oral and genital cavity. A diagnosis of Stevens Johnson’s syndrome was made. The patient had a history of cat I pulmonary TB and treated for 8 months and at the end of 8th month she was sputum smear negative. Four months later she had a relapse of sputum smear positive for pulmonary TB. She responded to the stoppage of drugs and oral/inj. Corticosteroids, antihistaminics and antibiotics.

Keywords: Rifampicin, Steven Johnson syndrome, Adverse drug reaction

ABSTRACT

A 25 year old female known case of category II pulmonary Tuberculosis was on anti-coch’s treatment in the FDC of rifampicin, isoniazid, pyrazinamide, ethambutol and Streptomyacin. Fifteen days after the commencement of Cat II anti TB treatment she developed diffuse erythematous rash on face, trunk and both extremities which turned into blisters. There were ulcers on oral and genital cavity. A diagnosis of Stevens Johnson’s syndrome was made. The patient had a history of cat I pulmonary TB and treated for 8 months and at the end of 8th month she was sputum smear negative. Four months later she had a relapse of sputum smear positive for pulmonary TB. She responded to the stoppage of drugs and oral/inj. Corticosteroids, antihistaminics and antibiotics.

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and started on ciprofloxacin 500mg twice daily, inj. cefotaxime 1gm i.v., chlorpheniramine 4mg twice daily, inj. dexamethasone 2mg i.v. then prednisolone 8mg and local lesions were symptomatically treated with fusidic acid & syp. aluminium hydroxide and magnesium hydroxide.

**DISCUSSION**

Steven Johnson Syndrome is a rare, immune complex hypersensitivity reaction characterized by sheet like skin and mucosal loss. It is potentially fatal with mortality rate of 5-15%. SJS has polyetiological pattern. The incubation period is typically a few days to 3 weeks but less than 48 hours in a patient with history of similar reaction to that drug. In this case the reaction occurred after 15 days of AKT. Over 100 drugs have been implicated. Most common drugs involved are cotrimoxazole, thiacetazone, phenylbutazone, oxicam group of drugs of NSAIDs, valdecoxib, carbamazepine, phenytoin, nevirapine, lamotrigine. SJS is also caused by viral; mycoplasma infection. There is no standard test to confirm the drug etiology. The causative agent is identified by history as no any confirmatory test methods available to detect the causative agent. Therefore identification of first event of ADR is based on the evaluation of probability. Here, the nature of Pharmacological agent and temporal association with clinical onset of the disease process is made.

Majority of cases begin with a week or two long prodrome of fever, malaise and nonspecific symptoms. This is followed by burning sensation, oedema, erythema of the lips and buccal mucosa and erythematous macules which rapidly get necrosed centrally with formation of vesicles, bullae and denudation of the face, trunk and extremities. Mucosal erosions occur in at least 2 sites – bulbar conjunctival, oral, nasal, anorectal junction, vulvovaginal region and urethra. Respiratory, GIT may get involved in complicated cases. The frequency in one review of 81 cases was oral mucosa 100%, eyes 91%, male genitalia 51% and anal mucous membrane 5%, while bronchitis and pneumonitis occurred in 6% and 23% of cases respectively.

The clinical differential diagnosis of SJS includes erythema multiform, viral exanthemas, ampicillin induced rash, morbilliform drug eruptions, Staphylococcal scalded skin syndrome, Kawasaki disease, acute graft versus host disease and SLE.

SJS has been known to be more common in patients with multisystem disorders, particularly those of collagen diseases. In our case when the patient had developed severe exfoliative dermatitis involving buccal, genital mucosa and severe conjunctivitis. All the anti TB drugs were stopped and the patient was treated with high doses of prednisolone and local steroids applied to eye and skin. She improved slowly after 3 weeks, once again the patient was considered good enough to have antitubercular drugs reintroduced one at a time. Rifampicin alone was started. But four days later she began to develop dermatitis, buccal ulceration and conjunctivitis similar to previous conditions. The rifampicin was promptly stopped and steroids were started again. She improved quickly and streptomycin, ethambutol, isoniazid, pyrazinamide were added stepwise with no further complication. The recurrence of SJS in our patient after the reintroduction of rifampicin strongly suggests a causal relation. A drug was defined as responsible for ADR if symptoms and signs resolved after withdrawal and recurred after rechallenge with that drugs. Attribution was also made if the cutaneous adverse drug reaction resolved with discontinuation of the drug, even with rechallenge.

So SJS requires a high index of suspicion by health personale. Knowledge about the condition and mode of treatment. Health personale should be extra vigilant who prescribe first line Anti TB drugs which are life saving and not be stopped without justification.

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