A case report: Stevens Johnson syndrome
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Introduction
Stevens johnson syndrome or erythema multiforme major is an acute, self limiting inflammatory condition of skin and mucous membrane. Here we report a case of a boy suffered from stevens johnson syndrome.

Case report
A 8 years old boy was admitted in pediatric ward, Dhaka Medical College Hospital (DMCH) with skin rashes and eruptions all over the body along with high grade continuous fever. This fever was started 3-4 days back and for this he was being treated with paracetamol and cotrimoxazole. After about 12 hours of starting the drugs, he developed this rash, which first appeared on the face and lips and then gradually spreaded all over the body. Along with this he also developed difficulties in swallowing and foreign body sensation in both eyes simultaneously. However, he had neither difficulties in micturition nor in defecation.Contour were normal. On further enquiry, mother also told that her child has been taking tegretol (carbamazepine) for last 14 days as prescribed for associated epilepsy.

On examination, the child was found febrile, temperature was 102o F, pulse 88/min, blood pressure 100/70 mm of Hg, maculo papular rashes were found all over the body specially over the extremities and trunk. Few lesions were characteristically pale centred with red surrounding ( "Target" lesions). Over the next 3 days the erythematous lesions became vesiculur and bullous and he developed ulcerations over the muco cutaneous junction of the lips and in the buccal mucosa with pseudomembrane formation. There was conjunctival congestion in both eyes. Anogenital mucosa was found healthy and urine color was normal.

Peripheral blood count shows total count of WBC- 17,000/cu mm, neutrophil-78%, lymphocyte- 20%, ESR-40 mm in 1st hour. Routine and microscopic examination of urine shows no abnormality, X-ray chest and ultrasonography of the whole abdomen reveals normal findings

At first the two suspected drugs (cotrimoxazole and carbamazepine) were withdrawn immediately. Then he was given supportive and symptomatic treatment with intravenous fluids, erythromycin, analgesics, prednisolone, glycerin swabs on lip, chloramphenicol eye drops and ointment. After about 48 hours, the child became stable; however, his treatment and other supportive care were continued for 10 days. Subsequently sodium valproate was started to prevent further epileptic attacks and also to see any untoward effects. Having no side effects of the drug and finding him clinically stable, the child was discharged after 15 days of hospitalization with advice to follow up and a cautionary advice regarding not to take cotrimoxazole and carbamazepine in future.

Discussion
Erythema multiforme is of two types minor and major, with erythema multiforme minor

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meaning skin lesions only, and erythema multiforme major being synonymous with stevens johnson syndrome.

Erythema multiforme minor as the name suggests is the less severe of the two types and accounts for 80% of erythema multiforme. The rash appears over a few days. There may be minor burning or itch. It is more intense over the backs of the hands and feet. The rash lasts for 1 to 2 weeks and then recedes leaving residual brown pigmentation.3

The hallmark of erythema multiforme major or stevens johnson syndrome is the development of large blisters in the mouth, on the skin, around the anus or genitals, in the throat or even on the eyes.3 In addition affected persons may develop reddish skin rashes in various shapes, sizes, and locations in addition to joint pains, fever and itching.

The majority of cases of stevens johnson syndrome are between ages 20 and 40, and 20% of cases occur in children and adolescents. It is rare both under the age of 3 and over the age of 50. Males are slightly more affected than females and there is no racial predilaction. The mortality of this syndrome is reported as 3 to 19%, although this number has probably come down with modern treatments.4

The most recognizable cause of stevens johnson syndrome is medications. If a person is prescribed a drug and develops a skin rash within two weeks, the cause is presumed to be the medication.3 Antibiotics are reported to cause at least 30% to 40% of cases, with sulfonamides, tetracyclines, amoxycillin, ampicillin and penicillin most commonly implicated. This syndrome occurs in less than 1 out of every 2000 people who take a form of penicillin, the most likely drug to cause this reaction.3 NSAIDs (butazones, ibuprofen, piroxicam and salicylates) and anticonvulsants (phenytoin, carbamazepine and phenobarbital) are also reported. Other possible etiologies include viral upper respiratory infections, mycoplasma pneumonia, pharyngitis, Herpes simplex infection, SLE, histoplasmosis, pregnancy, malignancy and external beam radiation. Some cases remain idiopathic. In our patient, the suspected drug is cotrimoxazole and carbamazepine is put in the 2nd position of the list as the patient had got it for two weeks.

The pathogenesis is unclear, but it may be a host specific cellmediated immune response to an antigenic stimulus, resulting in damage to keratinocytes cytokines released by activated mononuclear cells and keratinocytes may contribute to epidermal cell death and constitutional symptoms.

The diagnostic criteria for stevens johnson syndrome is individual "target" skin lesions less than 3 cm. in diameter, involvement of less than 20% of the body surface area, involvement of at least two mucous membranes namely the eyes, oral cavity, upper airway or esophagus, gastrointestinal tract or anogenital mucosa and biopsy compatible with this syndrome. The appearance of mucosal lesions is erythema and edema, which progresses to erosions and pseudomembrane formation. Stomatitis may be found in 100% of patients, ocular involvement in 86%, genital mucosal or urethral involvement in 41% and anal mucosal involvement in only 3% of patients. Upper airway mucosal involvement and pneumonia may be seen in up to 30% of cases. In our patient there was involvement of lips, oral cavity and eyes. In addition, virtually all patient will develop a characteristic maculopapular rash, usually early in the disease.

Prodromal symptoms, such as fever, malaise and cough are sometimes reported as a feature and they usually occur seven to ten days prior to fullblown presentation. Pain from mucosal ulceration is often severe, skin tenderness is minimal to absent. Corneal ulceration, anterior uveitis, panophthalmitis, bronchitis, pneumonia, myocarditis, hepatitis, enterocolitis, polyarthritis, hematuria and acute tubular necrosis leading to renal failure may occur.3
Other mucocutaneous syndromes which need to be excluded include Kawasaki's disease, Bechet's syndrome, small-vessel vasculitis syndromes, lupus erythematosus, pemphigus, pemphigoid, epidermolysis bullosa and dermatitis herpetiformis.

Management of stevens johnson syndrome is supportive and symptomatic. Prompt diagnosis, identification and early withdrawal of all suspected drugs are the most important preliminaries. Ophthalmologic consultation is mandatory. The management of the patients must be undertaken in specialized intensive care units with the same main types of therapy as for burns: warming of environment, correction of electrolyte disturbances, administration of a high caloric enteral intake and prevention of sepsis with appropriate antibiotics. Oral lesions should be managed with hydrogen peroxide mouth washes and glycerin swabs. Analgesics, antipruritics can also be used to relieve the annoying symptoms. The one longstanding controversy in the treatment of stevens johnson syndrome has been the use of corticosteroids. Although it is used in cases of life threatening systemic impairment, several reviews have concluded that steroids do not shorten the disease course and they produce more medical complications, namely secondary infections.

After healing follow-up is needed for ophthalmologic and mucous membrane sequelae such as ocular scarring and visual impairment, stricture of esophagus, bronchi, vagina, urethra or anus. Avoidance of offending drug and chemically related compounds is essential for the patient and first degree relatives.

References