Proceedings S.Z.P.G.M.I vol: 13(1-2) 1999, pp. 27-30.

Case Reports

Stevens Johnson Syndrome - A Case Report

Saba Abdul Sattar, Akbar Malik, Attiya Mahboob, Jehangir Durrani, Sajid Maqbool
Departments of Paediatrsc, Medicine and Ophthalmology, Shaikh Zayed Hospital, Lahore

INTRODUCTION

Stevens Johnson syndrome is the severe form of Erythema Multiforme, that's why it is also called Erythema Multiforme Major. It is a serious systemic disorder in which at least two mucous membranes and the skin are involved.

Incidence of Stevens Johnson Syndrome is about 0.8/million/year[1]. It is more common in children and young adults. No racial or sex preponderance is present.

CASE REPORT

Case - 1

A six year old child was admitted with the presenting complaints of; fever for 15 days, rash and watering of eyes for 4 days, inability to take food for 2 days.

According to the mother, the child was allright 15 days back when he developed high grade, intermittent fever not associated with chills and rigors. He was treated by a local G.P. who gave him Syrup Amoxil for 5 days and single dose of Fansimef Tablet.

Fever was temporarily relieved but one week later, mother noticed the appearance of rash over face, neck, ears and limbs with symmetrical involvement of palms and soles. The child also complained of watering of eyes with redness. Two days later he developed mouth ulceration and was unable to eat or drink.

History of weight loss, decreased appetite, headache and photophobia were present but there was no history of joint pains, palpitations, chest pain, diarrhoea or urinary complaints.

His vaccination was complete. Past history, birth history and developmental history was not significant. He was the student of Class-I and missing school because of his illness.

on examination

Temperature 101°F, respiratory rate 28/min, heart rate 110/mi, blood pressure 95/70 mmHg was recorded.

There was no jaundice, pallor or lymphadenopathy. A maculo papular rash was present over face, neck and trunk extending upto extensor surfaces of limbs. Lips were ulcerated and crusted with extension of lesions in oral cavity. Some oozing vesicles were present around the pinna. Eyes showed redness with yellow discharge. Lids, pupil and fundi were normal. Palms showed maculopapular as well as vesicobullous lesion extending upto forearms. Genetalia were not effected.

Investigations

Hb, 12 gm/dl, platelets 250,000, TLC 20.5, DLC P: 60%, L: 38%, E: 2%. Biochemistry values including serum electrolytes, BUN and serum creatinine were within normal range for his age. No pus cells/RBC present in urine. Blood culture showed no growth. Skin lesion scraping showed no growth. X-ray chest, ECG and Echocardiography were normal

Case - 2

This ten year old girl presented with complaints of fever for 5 days, rash for 2 days, redness of eyes and mouth ulcers for 2 days.

According to the mother, child had an episode of fit 2 weeks back, for which she was hospitalized. EEG revealed the presence of epileptic focus and she was put on carbamazepine one week back. She developed high grade fever five days before coming to hospital which was treated by a local G.P., with syrup Brufen and Augmentin. Three days back she suddenly developed rash all over the body, with watering of eyes and mouth ulceration.

History of weight loss marked photophobia and burning micturation were present. There was no history of joint pains, palpitations, chest pain or abdominal complaints..

She was fully vaccinated. Past history, birth and developmental history and family history was not significant. She was a student of Class-IV and could not go to school for 2 weeks because of her illness.

On examination she was very irritable, sick looking with temperature of 104°F, respiratory rate of 24/min, heart rate of 110/min and blood pressure of 100/70 mmHg. There was no jaundice, pallor or lymphadenopathy.

Face, neck, trunk and extremities showed maculopapular rash, out of which some lesions showed crusting. Ruptured bullae were also present over the chest and below the eyes. Lips were ulcerated with hemorrhagic - crusts with lesion extending into the oral cavity. A maculopapular rash with ruptured bullae and erythema was present over palms. **Systemic examination** was unremarkable and genitalia were also normal.

Investigations

- Hb 12 gm/dl, platelets 160,000, TLC 6.8, DLC P: 74%, L: 25%, E: 1%. Biochemistry values including serum electrolytes, BUN and serum creatinine were within normal range for his age.

No pus cells/RBC present in urine. Blood culture showed no growth. Skin lesion scrapping showed no growth. X-ray chest, ECG and Echocardiography were normal.

Diagnosis was of Stevens Johnson syndrome in both of these cases.

Sevens Johnson syndrome should be differentiated from:

1. **Kawasaki's Disease**
   It occurs in childhood with acute presentation just like Steven's Johnson syndrome and five criteria[2] should be met for its diagnosis. These include fever persisting for five or more days, changes of peripheral extremities starting with redness of palms and soles and later desquamation, conjunctival congestion, changes of lips and oral cavity with strawberry tongue and acute non-purulent cervical lymphadenopathy. Cardiovascular involvement with cardiomegaly, heart murmurs and ECG changes with prolonged PR and QT intervals and abnormal Q waves are important features of Kawasaki's disease.

2. **Eczema herpeticum (Kaposi varicelliform erruption)**
   It results from widespread infection of the eczematous skin with HSV[3]. Vesicles develop in large number abruptly over the areas of eczematous skin. Other parts of body like eyes, oral cavity and brain can also be affected by HSV with severe generalized spread of infection.

3. **Varicella**
   Caused by varicella zoster virus which spreads by droplet infection or through contact with Herpes zoster patient. The eruption appears over the trunk and then spread to face and limbs and axillae are almost always affected. Morbiliform erythema is followed by papules and tense clear vesicles rapidly changing to pustules. Vesicles in mouth are common and occasionally other mucous membranes like conjunctive may also be affected.

4. **Bullous Pemphigoid Juvenilis**
   It is a self limiting condition which appears is below six year age group[4]. Crops of tense bullae which may be blood stained occur on normal or etythematous skin especially over the face genitourinary regim and limbs. Mucous membranes may be affected.

5. **Staphylococcal Scalded Skin Syndrome**
   It is a blistering disorder caused by an epidermolytic toxin of certain phage type of Staphylococcus aureus. This condition is common in children[5] rapidly expanding, very shallow blisters which quickly rupture, leaving painful raw areas. All patients need anti-Staphylococcal antibiotics for cure.

Final Diagnosis

Based on history and examination supported by investigations, diagnosis in both of these cases was Stevens Johnsons Syndrome probably secondary to drug intake.

Treatment

Both cases were treated symptomatically with nutritional support, fluid and electrolyte imbalance was corrected by giving I/V fluid. Cleaning of skin was done with normal saline. To prevent secondary bacterial infection, antibiotics were used in the second case. Ophthalmological consultation was taken to prevent any eye complication.

Outcome

Both of the cases presented were successfully treated and discharged by giving symptomatic and supportive treatment.

DISCUSSION

Stevens Johnson Syndrme is an acute self limited systemic disorder, characterized by severe involvement of skin and more than one mucosal surfaces along with constitutional symptoms.

Incidence is 0.8/million/year. Individuals of all ages may be affected. No racial or sex preponderance present but it is more common in children and young adults[6].

Etiology

Various etiological factors include;

I) Infections;
- Viruses e.g. HSV, Adenovirus, Coxsackie virus.
- Bacteria e.g. Streptococcus, Staphylococcus, Proteus, Salmonella.
- Mycoplasma infections e.g. M. Pneumonae.
- Parasites e.g. Trichomonas
- Fungal infections e.g. Histoplasmosis

II) Ingestants;
- Food additives and dyes.
- Drugs: Many drugs can cause Stevens Johnson Syndrome but important ones, with which more cases are reported include; Sulfonamides, Penicillins, Barbiturates, Hydantoin, NSAIDS, Carbamezipine, Allopurinal.

III) Skin contactants e.g. creams containing sulfonamide, tropical wood etc.

IV) Physical factors like deep x-ray therapy, sunlight.

V) Malignancies e.g. lymphoma and leukemia can also be etiological agents.

VI) Miscellaneous group includes hormonal upset e.g. in pregnancy.

Pathogenesis of S.J. Syndrome

Pathogenesis is unknown[7] but immune mechanism involvement is suggested. Immunoglobulins and complement in and around the walls of dermal blood vessels with circulating immune complexes favours this hypothesis[8].

Clinical Features

Generally sudden in onset, history of drug intake or preceding viral infections may be present. Constitutional symptoms like fever, malaise, headache, rhinitis, myalgia and arthralgia is usually present.

Skin rash arises abruptly with symmetrical involvement[9]. Lesions appear first acrally and then in centripetal manner involving face, neck, extremities palms and soles. Prototype lesion is target lesion i.e. initially a macule is formed which enlarges, periphery is erythematous while centre becomes clear forming TARGET or IRIS lesion. Mucosal surfaces involvement is always present. Ulceration[10] and crusting of lips extends into the oral cavity affecting buccal mucosa, palate, tongue and extending upto larynx and oesophagus is seen.

Eye involvement occurs in most cases of Steven's Johnson syndrome[11]. The severity of involvement parallels that of lesions elsewhere, varying from mild hyperaemia to marked bullae formation and ulceration. Secondary infection may occur as a result of ruptured conjunctival bullae. Healing may be accompanied by subepithelial fibrosis, symblephron formation and kerato-conjunctivitis sicca, all of which may contribute to corneal opacification and vascularization with loss of vision.

Genitalia show purulent hemorrhagic inflammation which involves glans penis in males and vagina in female.

Systemic signs include persistent cough, vomiting, diarrhoa, abdominal pain, melena, hepatitis and myocarditis.

Complications of Stevens John Syndrome

- **Early:** Secondary bacterial infection; weight loss, anemia, dehydration and electrolyte imbalance leading to confusion, coma and seizures.
- **Late:**
- Eye problems e.g. trichiasis, corneal opacities and permanent, blindness.
- Skin may show scarring with contracture formation and alopecia.
- Stricture formation at various sites like oesophagus, branches, vagina and urethra.

Course and Prognosis

It varies from patient to patient. Usually it takes 3-6 weeks depending on severity and extent of

lesions. Minor forms will head in 10-14 days but S.J. syndrome may last for 8 weeks[1].

Prognosis is usually good. Mortality in severe form of disease is between 5-15%.

Management of S.J. Syndrome

It is supportive and symptomatic. Principles of management include;

1. Hospital admission.
2. Elimination of any suspected etiological factor if known.
3. Fluid and electrolyte imbalance correction by giving proper intravenous fluids.
4. Nutritional support.
5. Prevention of secondary bacterial infection to decrease mortality.
6. Ophthalmological consultations is mandatory to prevent eye complications as it can lead to permanent blindness.
7. Role of steroid in management of S.J. syndrome is controversial.

These should not be used routinely in EM major in general, but only under special circumstances and with particular caution. They may be justification in the very early stages of drug induced S.J. syndrome for 2-3 days or less in doses (80-180 mg/day) if disease progression has ceased. Doses ought to be stopped or tapered quickly but cautiously since no further benefit can be expected and the untoward effects may predominate thereafter.

REFERENCES

1. Peter O. Fritsch and Peter M. Elins. Erythema multiform and txic epidermal necrolysis. In: Dermatology in General Medicine (Thomas B. Fitzpatrick. Arthur Z. Eisen, Klaus Woll, eds.), 4th edition 1993; pp. 592-95. McGraw Hill. Inc. New York.
2. Robert R. Wolfe, MarcBoucek, Michael S. Schaffer James W. Wiggins. Mucotaneous lymph node syndrome. In: Current paediatric diagnosis and treatment (William W. Hay, Jassic R. Groothuis, Anthony, Myron, eds.) 13th edition, 1998; pp. 524. Prentice Hall Internal Inc.
3. McKee PH. Pathology of the skin-with clinical correlations (2nd edition). 1996. London: Times Mirror International Publishers Limited.
4. Donmokkes, Arnold. Odom. Andrew's diseases of the skin. In: Clinical dermatology, 7th edition, pp. 150-51. Igaku Shoin / Saunders International Edition.
5. Mackie RM. Clinical dermatology (4th edition). Oxford care texts. 1998. Oxford University Press, New York.
6. Gary L, Darm Stadt, Al Lane. Vesiculobullous disorders. Nelson text book of Paediatrics. 15th ed. pp. 1851-1852.
7. Bushkell LL, et al. Erythema multiforme: direct immunofluorescence studies and detection of circulating immune complexes. J Invest Dermatol 1980; 74: 372.
8. Huff JC, et al. Mixed cryoglobulinemia ^{125}I Clq binding and skin immunofluorescence in erythema multiforme. J Invest Dermatol 1980; 74: 375.
9. O. Braun Falcos, Plewing G, Wolf HH. Erythematous skin disorders. In: Dermatology, 3rd edition, 1984; pp. 409-12.
10. Harper J, Wells RS. Urticaria and erythemas. In: Handbook of Paeds dermatology 1985; pp. 80. Butterowrths.
11. Jack J, Kanski. Diseases of skin and mucous newborn. In: The eye in systemic diseases, 1986; pp. 113-114. Butterworth.

The Authors:

Saba Abdul Sattar
Tranee Registerar
Department of Paediatrics
Shaikh Zayed Hospital
Lahore

Akbar Malik
Assistant Professor
Department of Paediatrics
Shaikh Zayed Hospital
Lahore

Attiya Mahboob
Assistant Professor
Department of Dermatology
Shaikh Zayed Hospital
Lahore

Jehangir Durrani
Professor
Department of Ophthalmology
Shaikh Zayed Hospital
Lahore

Sajid Maqbool
Professor
Department of Paediatrics
Shaikh Zayed Hospital
Lahore

Address for Correspondence:

Saba Abdul Sattar
Tranee Registerar
Department of Paediatrics
Shaikh Zayed Hospital
Lahore