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STEVENS-JOHNSON SYNDROME: A CASE REPORT AND LITERATURE REVIEW

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ABSTRACT

The Stevens-Johnson disorder resembles a fractional thickness burns that may cause a 100% loss of epidermis, requiring a regeneration as a severe burn.[1] The case of interest was 62 years old female who is suffering from diabetes mellitus, hypertension, and pulmonary edema. She was referred from Al-Ahad general hospital to King Fahad General Hospital, Jizan, Saudi Arabia. She admitted there because of chest pain and tightness of breath then she developed oral ulceration and erythematosus skin patches and macules with erosions over the back. The treatment was planned as follow, applying Fucidin cream over eroded areas and fusiform over erythromatory areas, Nystatin 1ml oral drops, Chlorhexidine mouth wash and cyclosporine antibiotic or Intravenous immunoglobulin after medical evaluation. After seven days of continuous follow up the dermatological staff successfully diagnosed the patient as complicated Stevens-Johnson syndrome nd they suggest the appropriate treatment scheme.

KEYWORDS: Stevens-Johnson syndrome, Erythema, Ulceration.

INTRODUCTION

Stevens-Johnson syndrome (SJS) is considered as a type of severe skin reaction.[2] The primary induced symptoms were fever and flu-like symptoms.[2] After few days the skin starts to blister and peel forming painful, raw areas.[2] Mucous membranes, such as the mouth, are also typically involved.[2] Complicationsinclude dehydration, sepsis, pneumonia,

and multiple organ failures.[2] The clinical findings, diagnostic steps and management options are shortly discussed as well.

Case presentation

Our case was 62 years old female who is suffering from diabetes mellitus, hypertension, and pulmonary edema. She was referred from Al-Ahad general hospital to King Fahad General Hospital, Jizan, Saudi Arabia. She admitted there because of chest pain and tightness of breath then she developed oral ulceration and erythematosus skin patches and macules with erosions over the back.

After the clinical examination, the following signs and symptoms were observed:

- Detachment body surface area (BSA) > or =25% especially appeared on the back, both small nodules of the breast and abdomen (Figure 1 and 2).
- Dusky erythema over upper limbs faced upper back (figure 2).
- Rashes all over the body and skin are peeling over the back.
- Crusted laceration over lesions over the lip, eyelid and ear.
- Mouth ulcer.
- Conjunctivitis.

Figure 1: Characteristic erythema and skin detachment in abdomen.

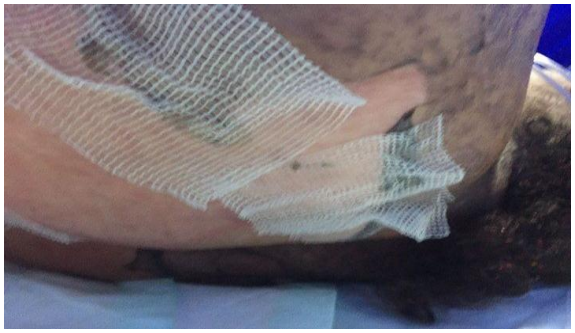

Figure 2: Characteristic erythema and skin detachment in back.

As for the patient's clinical course, the vitality measures were of concern, and its results were as follows

- The body temperature never above 37°C during the subsequent seven days of hospitalization.
- Cytosolic blood pressure: 90 – 154 mmHg.
- Diastolic blood pressure: 80 – 57 mmHg (i.e., good controlled blood pressure).

Laboratory investigations during the seven hospitalization days have the following results

- White blood cells count (WBCs): 5,300-9,900 cell/cm^3.
- Hemoglobin (Hb%): 8.1 – 9.9 gm/dl.
- Normal platelets count.
- Normal kidney function tests.
- Liver function tests were slightly elevated on the first day of hospitalization (AL= 53 and AST= 82), after that the liver function was normal.
- Normal blood electrolytes level (Sodium, Phosphorus, Potassium, and Magnesium).
- On the first day, the Erythrocyte Sedimentation Rate (ESR) was hight (116 mm).
- Normal coagulation profile (PT and PTT).
- Lactate dehydrogenase: 272 – 353 U/L.
- Normal alkaline phosphatase level.
- Mildly elevated blood glucose level.
- Blood culture results were negative for aerobic and non-aerobic bacteria.
- Eye culture and sensitivity give a positive result for staphyl. aures MRSA and it reported that she was resist for aminoglycosides, carbapenemes, quinolones, macrolides, penicillin and combinations, sulfonamides and cephalosporin.
- Normal arterial blood gases.

Radiological scanning during hospitalization days

• Chest X-ray scans: Mild obliteration left costophrenic angle.

The ocular examination was as follows

- Skin ulceration of upper and lower lids in both eye.
- Congested Conjunctivitis.
- Clear cornea in both eyes.

The patient was planned to be treated as follows

- Firstly the patient should be seen by medical as she has diabetes mellitus, hypertension, pulmonary edema and chest pain.
- Applying Fucidin cream over eroded areas and Fusicort over erythromatory areas.
- For possible Methylpyridizone, cyclosporine or intravenous immunoglobulin (IV/IG).
- Start intravenous Methylpralnbondone 40 mg IV OD.
- Nystatin 1 ml oral drops.
- Chlorhexidine mouth wash.
- For eyes: continue topical prednisolone + Ciprofloxacin + Lubricant eye drops.
- Continues daily dressing BID.

According to the recommendations of dermatological clinic the patient was finally advised for

- Acyclovir 200 mg tablets (400) (orally) (Q6H).
- 7 prednisolone (Gupisone) 5 mg tablet (5) (orally) (daily).
- 10 Vaseline local apply 40 gm (10) (topical) (BID).
- 90 Fusidic acid 2% (20 mg/g) ointment (1) (topical) (BID).
- 7 Glibenclamide 5 mg tablet (2.5) (orally) (daily).
- 9.0 Lactulise 10 g/15 ml in 300 ml syrup (10) (orally) (TID).

After seven days of hospitalization and continuous follow up, the patient completely recovered and left the hospital and is in good health.

DISCUSSION

Stevens-Johnson disorder (SJS) and toxic epidermal necrolysis (TEN) are uncommon but dangerous mucocutaneous diseases that dominatingly happen as unfavorable responses to recently directed medications. A few past observational examinations distinguished antiepileptics, allopurinol, and certain antimicrobials (sulphonamide anti-toxins are the major ones) as those drugs have the highest risk for causing SJS/TEN. However other medications have been related with less evidence (e.g., oxicam analgesics, sertraline [a specific serotonin reuptake inhibitor, or SSRI], COX-2 inhibitors).[3]

Epidemiology

SJS and TEN are uncommon diseases with an occurrence of 1.89 instances of TEN for each million occupants for each year reported for Western Germany and Berlin in 1996.[4] La

Grenade et al report comparative outcomes, with 1.9 instances of TEN for every million occupants for every year given all cases answered to the FDA AERS database in the USA.[5] Lower incidence was reported by Chan et al. in Singapore.[6] Certain conditions may affect the frequency of TEN, and this is plainly the case for HIV where the yearly incidence is around one thousand times higher than in the all-inclusive community, with roughly 1 case for each thousand every year in the HIV-positive populace.[7]

Signs and symptoms

SJS ordinarily starts with fever, sore throat and fatigue, **which is commonly misdiagnosed and consequently treated with** antibiotics. SJS and TEN are often heralded by fever, sore throat, cough, and burning eyes for 1 to 3 days. Patients with SJS and TEN frequently experience the burning pain of their skin at the start of disease.[8] Ulcers and other lesions begin to appear in the mucous membranes, almost always in the mouth and lips, but also in the genital and anal regions. Those in the mouth are usually extremely painful and reduce the patient's ability to eat or drink. Conjunctivitis of the eyes occurs in about 30% of children who develop SJS]. A rash of round lesions about an inch across arises on the face, trunk, arms and legs, and soles of the feet, but usually not the scalp.[9]

Etiology

Different etiologic variables have been identified as a cause for Stevens-Johnson disorder. Medications most ordinarily are faulted. The four etiologic categories are as follows:

- Infectious
- Drug-induced
- Malignancy-related
- Idiopathic

Stevens-Johnson syndrome is idiopathic in 25-50% of cases. Drugs and malignancies are most often implicated as the etiology in adults and elderly persons. Pediatric cases are related more often to infections.

Infectious causes

Viral diseases that have been reported to cause Stevens-Johnson syndrome include the following:

- Herpes simplex virus (possibly; remains a debated issue)
- AIDS

- Coxsackie viral infections
- Influenza
- Hepatitis
- Mumps

Bacterial etiologies include the following:

- Group A beta-hemolytic streptococci
- Diphtheria
- Brucellosis
- Lymphogranuloma venereum
- Mycobacteria

Mycoplasma pneumonia.[10,11]

- Rickettsial infections
- Tularemia
- Typhoid

Possible fungal causes include coccidioidomycosis, dermatophytosis, and histoplasmosis. Malaria and trichomoniasis have been reported as protozoal causes.

Drug-induced

Antibiotics are the most common cause of Stevens-Johnson syndrome, followed by analgesics, cough and cold medication, NSAIDs, psycholeptics, and antigout drugs. Of antibiotics, penicillins and sulfa drugs are prominent culprits; ciprofloxacin has also been reported.[12]

The following anticonvulsants have been implicated

- Phenytoin
- Carbamazepine
- oxcarbazepine (Trileptal)
- Valproic acid
- Lamotrigine
- Barbiturates

Stevens-Johnson syndrome has additionally been diagnosed in patients taking the following medications

- Modafinil (Provigil)
- Allopurinol.[13]
- Mirtazapine.[14]
- TNF-alpha antagonists (eg, infliximab, etanercept, adalimumab).[15]
- Cocaine
- Sertraline
- Pantoprazole
- Tramadol

Genetic factors

The presence of the following human leukocyte antigens genes has been associated with increased risk of Stevens-Johnson syndrome:

- HLA-B*1502
- HLA-B*5801
- HLA-B*44
- HLA-A29
- HLA-B12
- HLA-DR7
- HLA-A2
- HLA-B*5801
- HLA-A*0206
- HLA-DQB1*0601

DIAGNOSIS

Diagnosis all suspected cases of SJS and TEN should be confirmed by skin biopsy for histologic and immunofluorescence examinations. Early lesion shows suprabasal layer apoptotic keratinocytes. Later lesion shows full-thickness epidermal necrosis and separation of epidermis from dermis. Some important conditions mimic SJS and TEN hence a histological evidence is important. Since 90% SJS and TEN have mucous membrane involvement the absence of such should prompt one to consider alternative diagnosis.[16]

TREATMENT

The essential treatment for SJS/TEN is the end of the causative factor(s), typically an offending medication. The speedier the causative medication is dispensed with, the better the prognosis.[17] Another imperative component of treatment that has been proposed is supportive care in a burn unit or intensive care.[17] There is some proof that cyclosporine, cyclophosphamide, plasmapheresis, fundamental corticosteroids, tumor necrosis factor-alpha (TNF-α) inhibitors, and IVIG might be valuable; but, treatment with foundational corticosteroids and other immunosuppressive drugs is controversial as a result of a possible high danger of sepsis.[17]

CONCLUSION

Stevens-Johnson syndrome (SJS) is a rare syndrome. Some medicines are related to a high danger of Stevens-Johnson syndrome. There is a strong association between genetics and reaction to some medication strong genetic associations. Early diagnosis plays a vital role in response to treatment. Continuous follow up is strongly requested.

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