

A CASE REPORT OF STEVEN JOHNSON SYNDROME

Abstract

Introduction: In 1922, two doctors, Albert Mason Stevens and Frank Johnson, examined purulent conjunctivitis.” **Background:** Stevens-Johnson syndrome was named after them as a result of their study. The incidence rate is 7 cases per million populations per year. **Case Presentation:** Master Yash Ghudam was brought to AVBRH by his parents with chief complaints of fever since 5 days and erythematous lesions all over body since 3 days. **History of present illness:** Patient was apparently alright 5 days back, and then he started having fever which was of high grade and was not associated with chills and rigor. Patient was treated on OPD basis and the symptoms of an unexplained disease in two young boys, aged 7 and 8, who had "an unusual, generalised eruption of continued fever, inflamed buccal mucosa, and extreme some antibiotic was given, but there was no relief, after 2 days there was ulcers formation inside the mouth for which some ointment and syrup becosule was started. But lesions were increasing. 3 days back the lesions first appeared on chest then got spread to legs and hands. For which patient was admitted in Chandrapur hospital from where the patient was referred to AVBRH for further management. **Interventions:** The patient was treated the patient was started on intravenous and orally Cortecosteroids, Omnacortil 10mg, Antibiotics- Inj. Ceftriaxone 1gm IV 12 hourly [100mg/kg/day], inj. Amikacin 150mg IV 12 hourly [15mg/kg/day], Syp. Mucaïne gel 2tsp BD – swish and swallow), Syp. Cital 2.5ml TDS, Tab. Chymoral Forte TDS, Inj. Pantop 20mg IV 24 hourly (1mg/kg/dose), **Pandya’s Formula-** Syp. Gelusil 5ml, Syp. Benadryl 5ml, Syp. Omnacortil 5ml. Skin allograft: It has been planned. **Conclusion:** In this study, we mainly focus on medical management and outstanding nursing care helped prevent further complication. Overall, the patient's reaction was positive, though recovery time from Steven Johnson syndrome varies from person to person, taking weeks, months, or even years. However, only a small number of people completely recover, while some have long-term consequences. She took a long time to get back on her feet. **Keyword :** stevenjohnson syndrome, fever, erythematous lesions.

Introduction

Stevens-Johnson syndrome is a hypersensitivity complex affecting the skin and mucous membranes that is caused by an immune complex. It can affect the mucous membranes of the mouth, nose, eyes, genitals, urethra, gastrointestinal tract, and lower respiratory tract. Necrosis may result from GI and respiratory involvement. Stevens-Johnson syndrome is a severe systemic condition that can cause substantial morbidity and mortality.

Difference between Stevens- Johnson syndrome and Toxic Epidermal Necrolysis:

Except for the delivery, Stevens-Johnson syndrome and toxic epidermal necrolysis are clinically identical. Changes affect less than 10% of body surface area in SJS and more than 30% of body surface area in TEN, according to one generally agreed definition; involvement of 15 to 30% of body surface area is called SJS/TEN overlap. 1. Small patches of peeling skin are caused by Stevens-Johnson syndrome (affecting less than 10 percent of the body). 2. Large areas of peeling skin are caused by toxic epidermal necrolysis (affecting over 30 percent of the body).

Classification of Stevens-Johnson syndrome: 1. Stevens-Johnson syndrome is a "minor type of TEN" characterised by a detachment of less than 10% of the body surface area. 2.Overlapping Stevens-Johnson syndrome/toxic epidermal necrolysis: body surface area detachment of 10-30%. 3.Toxic epidermal necrolysis: More than 30% of the body's surface area is detached.

Case history

Patient information

Master Yash Ghudam was brought to AVBRH by his parents with chief complaints of fever since 5 days and erythematous lesions all over body since 3 days. Patient was apparently alright 5 days back, and then he started having fever which was of high grade and was not associated with chills and rigor. Patient was treated on OPD basis and some antibiotic was given, but there was no relief, after 2 days there was ulcers formation inside the mouth for which some ointment and syrup becosule was started. But lesions were increasing. 3 days back the lesions first appeared on chest then got spread to legs and hands. For which patient was admitted in Chandrapur hospital from where the patient was referred to AVBRH for further management.

Medical/Surgical History:There is no significant history of any medical and surgical illness in past.

Psychosocial history: Patient maintains good interpersonal relationship with family, friends and relatives.

Family history: Mr. UmeshGhudam is a known case of diabetes mellitus and hypertension. There is no other family history of illness.

Socioeconomic status: Mr. UmeshGhudam is the only bread winner of the family he is electrician and works in MSEB he earns 25,000/- month approximately. They live in their own house made up of bricks and cement. There is proper electricity and water supply in their house.

Nutritional status: They consume mixed diet.

Personal history: Patient is having disturbed sleeping pattern. Hygiene is well maintained.

Physical examination:

General parameter: Temperature- 101⁰F, Pulse- 120 breath/ min, Respiration-26 breath/ min, Heart sound- 118 beats/min, Blood pressure- 110/70mmhg.

Height- 130cm, Weight – 20kg, Head circumference -53cm, Chest circumference – 55cm , Mid Arm circumference – 16cm, BMI – body mass index = 11.8 /M2, 14 percent Underweight. Healthy BMI range- 14to 18.6kg/m2.

Mental status: He was conscious and he had a Glasgow Coma Scale of 15 given the response after the stimulation. **Skin condition:** Skin colour- whitish, Pigmentation- present, Lesion- present, Cyanosis – not present, Scar – present, Oedema – present, Tenderness – not present, Rash – present. **Face:** Expression - dull, Symmetry – symmetrical, Tenderness over Sinuses – not present, sinuses- not present. **Eyes: Periorbital edema-** present- due ti inflammation, **Photophobia-** not present, Pupils- equally reacting to light, conjunctiva- present congestion.

Mouth And Throat: Colour of lips- Whitish, Gum bleeding- Pink, Swelling-present, lesions of buccal- present, Mucosa-Present Tonsillitis- not present. **Neck:** Range of motion- Normal range of motion present, Lymph node- No enlargement. **Chest: Inspection-** Lesion, scar, **Palpation-** No auxiliary lymph node, **Percussion-** Plural effusion absent, **Auscultation-** S1 and S2 murmur sound herd.

Diagnosis assessment : Physical examination and history collection: Based on the patient's medical history and a physical examination, diagnose Stevens-Johnson syndrome. **Skin biopsy:** - A sample of skin can be removed for laboratory testing to confirm the diagnosis and rule out other potential causes (biopsy). **Culture test:** - Skin or oral culture or culture from other areas may be taken to confirm or rule out infection - (In my patient culture test are done by antibiotic sensitivity test) Amikacin: Resistant, Ceftazidim: -Resistant , Ciprofloxacin:- sensitive, Tetracycline:- sensitive, Cotrimoxazole:-Resistant, Gentamycin:-Resistant, Colistin:- sensitive. **Imaging:** - Depending on symptoms, a chest X-ray to check for pneumonia. **Blood tests:** These are used to rule out the risk of infection or other causes.

Investigation: CBC investigation

Hb%- 9.8gram, MCHC- 32.4g/dl, MCV- 61cub. Micron, MCH- 41.3, Total RBC count- 4.99 million cell per microliter, Total WBC count- 8000microliter, Total Platelet count- 5.53, HCT- 30.3%, Monocytes – 02%, Granulocytes- 63%, Lymphocytes- 33%, RDW- 13.3 %k/ul, Eosinophils- 0.2%, Basophils- 00 k/ul. **Kidney Function Test-** Urea- 22mg/dl, Creatinine- 0.4 milligram, Sodium- 127 meg/l, Potassium- 4.9 meg/l, **Liver Function Test-** Alkaline phosphatase – 70 IU, ALT (SGPT)- 20 IU, AST (SGOT)- 32 IU, Total protein- 6.5 IU, Albumin- 2.8 G/dl, Total bilirubin- 0.6mg/dl, Conjugated bilirubin- 0.4mg/dl, Unconjugated bilirubin- 1.1 mg/dl, Globulin calculated- 3.7 mg/dl, **CRP-** 10.6 mg/dl.

Therapeutic intervention

General measures: To check the vital sign (Temperature pulse respiration and BP.) airway, fluid and electrolyte balance and prevention of complications like seizures, pulmonary aspiration, pressure source, thrombophlebitis are mandatory. Health management includes physiotherapy, health diet.

Management:

The majority of Stevens-Johnson syndrome patients are treated in intensive care units or burn centres. Since there is no particular treatment for Stevens-Johnson syndrome, most patients are treated symptomatically.

Stopping nonessential medications

To cure Stevens-Johnson syndrome, the first and most significant step is to avoid taking any drugs that may be causing it. Since it's difficult to pinpoint which drug is causing the problem, your doctor can advise you to discontinue all non-essential medications.

Supportive care

If you're in the hospital, you're likely to receive the following forms of supportive care:

1. **Aseptic precautions:** Strict aseptic precautions have to be maintained to prevent further infection and complications.
2. **Fluid replacement and nutrition:** Since skin loss may cause a substantial loss of fluid in your body, fluid replacement is a necessary part of treatment. A tube inserted through your nose and advanced through your stomach may be used to administer fluids and nutrients to the patient (nasogastric tube). - The patient is free to eat and drink. -IV fluid (100%) DNS- 500ml 8hourly.
3. **Wound care:** Blisters can be soothed with cold, wet compresses as they recover. Your doctor can gently remove any dead skin and cover the affected areas with petroleum jelly (Vaseline) or a medicated dressing. Burns are treated as skin lesions. Saline compresses should be applied to areas of denuded skin.
4. **Eye care:** A patient can also need the services of an eye specialist (ophthalmologist). A aggressive lubrication of the ocular surface is typically the first step in treating acute ocular manifestations. Most ophthalmologists treat inflammation and cicatric changes with topical steroids, antibiotics, and symblepharonlysis. **Symblepharonlysis:** A symblepharon is a partial or total adhesion of the palpebral conjunctiva of the eyelid to the bulbar conjunctiva of the eyeball, which is normally broken down by mechanical means. It's triggered by an illness or a traumatic incident.

Medical management:

IVIg - Given (**Human normal immunoglobulin**10gm in 100ml solution over 3 hrs.)-

Instructions: Mid BT Lasix 20 mg IV stat, In case of any adverse reaction stop transfusion immediately. Inj. Avil 4 mg stat, Inj. Hydrocortisone 100 mg IV start.

Corticosteroids- Given Tab. Omnacortil 10mg 2---0,

Antibiotics Given -

1. Inj. Ceftriaxone 1gm IV 12 hourly [100mg/kg/day]
2. Inj. Amikacin 150mg IV 12 hourly [15mg/kg/day]

Syp. Mucaine gel - Manage oral lesions with mouthwashes. Topical anaesthetics are useful in reducing pain and allowing the patient to take in fluids.- Given (Syp. Mucaine gel 2tsp BD – swish and swallow).

Others - Syp. Cital 2.5ml TDS , Tab. Chymoral Forte TDS, Inj. Pantop 20mg IV 24 hourly (1mg/kg/dose).

IVIg: Immunoglobulin therapy, also known as natural human immunoglobulin (NHIG), is a treatment that uses a combination of antibodies (immunoglobulins) to treat a variety of illnesses. These include primary immunodeficiency, immune thrombocytopenic purpura, chronic inflammatory demyelinating polyneuropathy, Kawasaki disease, some cases of HIV/AIDS and measles, Guillain-Barré syndrome, and some other conditions. It may be injected into a muscle, a vein, or under the skin, depending on the formulation. The effects last for a couple of weeks.

Inj Lasix: Furosemide (Lasix) is a diuretic (water pill) that helps the body rid itself of excess water and salt. Salt (sodium and chloride), water, and other small molecules are usually filtered out of the blood and into the tubules of the kidney by the kidneys. Urine is formed from the filtered solution. The majority of the sodium, chloride, and water washed out of the blood is reabsorbed. Furosemide functions by preventing sodium, chloride, and water from the filtered fluid in the kidney tubules from being absorbed, resulting in a significant increase in urine production (diuresis). The diuresis lasts about 6-8 hours after oral administration, and the onset of action is within one hour. The operation begins five minutes after injection, and the diuresis lasts two hours. Furosemide's diuretic effect can lead to a loss of sodium, chloride, body water, and other minerals.

Inj. Avil: Avil contains pheniramine maleate, an antihistamine used to treat hayfever, runny nose, itchy skin, and skin rashes. It's also used to avoid and treat inner ear conditions like

Meniere's disease, as well as travel sickness. Avil belongs to a class of drugs known as 'antihistamines,' which function by inhibiting the action of histamine. It belongs to the alkylamine family of antihistamines and is a first-generation antihistamine. It competes with histamine for the H1 receptor and, once bound, acts as an inverse agonist.

Inj. Hydrocortisone: Medicinal Hydrocortisone is a synthetic or semisynthetic derivative of natural hydrocortisone hormone, which is formed by the adrenal glands and has glucocorticoid and mild mineralocorticoid effects. Hydrocortisone promotes protein catabolism, gluconeogenesis, capillary wall stabilization, renal calcium excretion, and suppresses immune and inflammatory responses by acting as a glucocorticoid receptor agonist. Corticosteroid Hormone Receptor Agonist is how hydrocortisone works.

Tab. Omnacortil : Prednisolone is a cortisol-like glucocorticoid used for its anti-inflammatory, immunosuppressive, anti-neoplastic and vasoconstrictive effects. Glucocorticoids inhibit apoptosis and demargination of neutrophils; Inhibits phospholipase A2, which reduces the formation of arachidonic acid derivatives; inhibits NF-Kappa B and other inflammatory transcription factors; promotes anti-inflammatory genes such as interleukin-10.

Inj. Ceftriaxone: A wide-spectrum cephalosporin antibiotic with a very long half-life and high penetrability to meninges, eyes and inner ears. Ceftriaxone is a cephalosporin/cephamycin beta-lactam antibiotic used to treat bacterial infections caused by susceptible, usually gram-positive, organisms. Ceftriaxone works by inhibiting the synthesis of mucopeptides in the bacterial cell wall. Ceftriaxone binds to carboxypeptidases, endopeptidases, and transpeptidases in the bacterial cytoplasmic membrane with its beta-lactam moiety.

Inj. Amikacin: Amikacin is an antibiotic that belongs to the aminoglycoside family. Aminoglycosides bind to bacteria, allowing t-RNA to be misread, preventing bacteria from synthesizing essential proteins. Aminoglycosides are mostly used to treat infections caused by Gram-negative aerobic bacteria like Pseudomonas, Acinetobacter, and Enterobacter. Amikacin's primary mechanism of action is the same as with other aminoglycosides. It binds to 30S ribosomal subunits in bacteria and interferes with mRNA binding and tRNA acceptor sites, halting bacterial development. As a result, normal protein synthesis is disrupted, and non-functional or toxic peptides are produced.

Syp. Mucaine gel: Mucaine suspension combines an antacid effect (Aluminum hydroxide and Magnesium hydroxide) with a topical local anesthetic (Oxethazaine) to relieve pain associated with hyperacidic conditions. To neutralize stomach acid, aluminium and magnesium hydroxide react. Oxethazaine is a potent local anesthetic that can be used in extremely small amounts. It numbs the stomach walls and relieves the pain associated with hyperacidic conditions in a variety of ways.

Syp. Cital: Disodium Hydrogen Citrate is a Systemic alkalizer that relieves the discomfort of cystitis caused by lower UTI.

Tab. Chymoral Forte: Chymotrypsin is a digestive enzyme produced in the pancreas that aids in the breakdown of proteins and polypeptides, a process known as proteolysis. Chymotrypsin is made in the pancreas as chymotrypsinogen, an inactive precursor that is secreted to the duodenum and triggered by trypsin-induced cleavage. It also activates itself by cleaving critical amino acid residues in the oxyanion hole to create -Chymotrypsin, which is more stable than -Chymotrypsin.

Inj. Pantop: Pantoprazole works by inhibiting the final step in the synthesis of gastric acid. Pantoprazole inhibits gastric acid and basal acid secretion by covalently binding to the H⁺/K⁺ ATP pump in the stomach's gastric parietal cell. For up to 24 hours and longer, the covalent binding inhibits acid secretion.

PANDYA'S FORMULA:

The Pandya's formula is a formula which is used for the treatment of erosion of mucous membrane due to disease condition such as Steven's Johnson Syndrome, Toxic Epidermal Necrolysis (TEN), etc. The formula contains three types of medications.

- Syp. Gelusil 5ml
- Syp. Benadryl 5ml
- Syp. Omnacortil 5ml

This mixture should be softly rinsed in the mouth for 2 mins and then spitted out. Since cutaneous erosions recover in 1–2 weeks, the treatment for them is mainly supportive.

- 1. SYP. GELUSIL: Composition:** Aluminium hydroxide, simethicone, magnesium
Action: Antacids made of aluminum or magnesium act easily to reduce stomach acid. Antacids in liquid form normally work quicker and stronger than antacids in tablet or capsule form. Simethicone aids in the breakdown of gas bubbles in the intestine. Heartburn, indigestion, bloating, and belching are symptoms. **Side effects:** Nausea, constipation, diarrhea, headache, loss of appetite, unusual tiredness, muscle weakness.
Contra-indication:
- 2. SYP. OMNACORTIL: Corticosteroid Composition:** Prednisolone Sodium Phosphate
Action: decreases inflammation by suppression of migration of polymorphonuclear leukocytes, fibroblasts; reversal to increase capillary permeability and lysosomal stabilization.
Indication: Severe inflammation, immunosuppression, neoplasms. **Side effects:** CNS depression, headache, hypertension, tachycardia, thrombophlebitis, increased intraocular pressure, blurred vision, GI haemorrhage, increased appetite. **Contraindication:** Children <2 years, psychosis, hypersensitivity, acute glomerulonephritis, measles, idiopathic thrombocytopenia.
- 3. SYP. BENADRYL: 1st generation antihistamine Composition:** Diphenhydramine
Action: Acts on blood vessels, GI, respiratory system by competing with histamine for H₁ receptor site, decreases allergic response by blocking histamine. **Indication:** Allergy symptoms, rhinitis, parkinsonism, nighttime sedation, infant colic, non-productive cough, insomnia in children. **Contra-indication:** Acute asthma attack, lower respiratory tract disease, and neonates are all linked to hypersensitivity to H₁ receptor antagonists. **Side effects:** Dizziness, poor coordination, fatigue, confusion, blurred vision, dilated pupils, chest tightness, urinary retention, dysuria, thrombocytopenia.

Surgical management:

Skin allograft: For patients with Steven Johnson syndrome, skin grafting with glycerol-preserved donor skin obtained from a skin bank is performed.

Oxygen therapy:- oxygen administration 2 liters/min through a nasal catheter.

Nursing management:-

First of all makes nursing assessment with the help of observation to check the consciousness, weakness, speech, vital sign, the reaction of a pupil, size of a pupil. To make the client lie comfortably in bed. After checking vital signs ensure patient airway and to given O₂ therapy .elevate head end of the bed to 30° and railing bed is provided. To monitor BP.

Nursing diagnosis

1. Impaired thermoregulation related to infection and inflammation secondary to steven johnson syndrome.

- **Short term goal:** To reduce body temperature to normal.
- **Long term goal:** To maintain normal body temperature.
- **INTERVENTION-** **1.** To monitor vital signs (to check variations in body temperature). **2.** To provide cool environment and proper ventilation (to maintain normal body temperature). **3.** Provide cold sponge bath if the temperature exceeds more than normal (to reduce body temperature to normal). **4.** To administer medications as prescribed by the physician (to treat the cause of fever and to reduce the body temperature to normal).

2. Acute pain related to inflammatory erythematous lesions secondary to steven johnson syndrome.

Short term goal: To reduce pain.

Long term goal: To minimize pain.

- **INTERVENTION:-** **1.** To monitor intensity, quality and duration of pain (to assess for pain level). **2.** Teach patients non-pharmacological techniques (e.g., relaxation techniques, divertional therapy, deep breathing exercises etc.), (helps to alleviate pain and provides comfort). **3.** Explore the factors that worsen pain (to avoid the factors from worsening the pain). **4.** Administer medications as prescribed by physician (to reduce pain).

3. Impaired skin integrity related to erythematous lesions all over the body secondary to steven johnson syndrome.

Short term goal: To maintain normal skin texture.

Long term goal: To maintain normal skin integrity.

- **INTERVENTION:-**1. Assess the overall condition of the skin (it aids in providing possible intervention related to skin integrity). 2. Evaluate the patients awareness of sensation of pressure (patients who are unaware of sensation tends to do nothing thus results in prolonged pressure on skin capillaries leading to ischemia).

Communication: Assess the difficulty in using language to communicate or question answer. Encourage the patient's effort to communicate. Speak slowly in simple and also provided consult speech therapists.

Conclusion: Master. Yash Ghudam was admitted in PICU with chief complaints of fever, rashes and erythematous for which he was treated on OPD basis and some antibiotic was given, but there was no relief, after 2 days there was ulcers formation inside the mouth for which some ointment and syrup becosule was started. But lesions were increasing. 3 days back the lesions first appeared on chest then got spread to legs and hands. For which patient was admitted in Chandrapur hospital from where the patient was referred to AVBRH for further management. He is undergoing treatment like IVIG, antibiotics cortecosteroids etc. skin allograft is planned for him.

Acknowledgement

The thanks to Dr.Seema Singh, Professor cum Principal, Smt.Radhibabai Meghe Memorial College of Nursing. Datta Meghe Institute of Medical Sciences, Sawangi (M) Wardha for her timely support and valuable suggestions.Mrs. Jaya Gawai, Asso.professor cum academic dean .Mrs.Vaishali Taksande, Professor Dept. of OBGY, The author also thanks Mrs. Archana Maurya, Professor, Dept. of Child health Nursing. Smt.Radhibabai Meghe Memorial College of Nursing, Datta Meghe Institute of Medical Sciences, (Deemed to be University), Sawangi (M) Wardha, for their continuous support and valuable suggestions. The Authors are also grateful to authors / editors / publishers of all those articles, journals and books, from where the literature for this article has been reviewed and discussed. Authors are grateful to IJCRR editorial board members and IJCRR team of reviewers who have helped to bring quality to this manuscript.

Ethical approval

Not applicable

Patient Inform consent

While preparing case report and for publication patient's informed consent has been taken.

Conflict of Interest there are no conflicts of interest.

Funding

The author declare that

Not applicable

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