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Idiopathic Stevens-Johnson Syndrome in a Child: a Case Report

Amir Hossein Goudarzian[1], Mahya Ghahramanlou[2], Sina Ehsani[3], Ali Abbas khanian[4], *Afshin Shourofi[5]

[1]BSc of Nursing, Student Research Committee, Mazandaran University of Medical Sciences, Sari, Iran. [2]Medical Student, Mazandaran University of Medical Sciences, Sari, Iran. [3]BSc of Radiology, Student Research Committee, Mazandaran University of Medical Sciences, Sari, Iran. [4]Department of Pediatric Neurology, School of Medicine, Mazandaran University of Medical Sciences, Sari, Iran. [5]Traditional & Complementary Medicine Research Center, Mazandaran University of Medical Sciences, Sari, Iran.

Abstract

Background

Stevens-Johnson syndrome (SJS) is one of potentially fatal disorders that often occur after taking certain types of medication. There are reports of this disease after some infections. This paper presents the case of a young boy with idiopathic Stevens-Johnson syndrome.

Case Report

A six-year-old male patient with complaint of fever and skin lesions was transferred to emergency department of Bu-Ali Sina hospital (Center of Mazandaran province, Iran). After further evaluations (Physical examination and laboratory reports), the patient did not show any common causes of Stevens - Johnson syndrome. Fortunately he was discharged in a good condition after duration of treatment.

Conclusion

Further studies need to be done in the field of risk factors of Stevens-Johnson syndrome.

Key Words: Child, Idiopathic, Case report, Iran, Steven-Johnson syndrome.

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Corresponding Author:

Dr. Afshin Shourofi., Traditional & Complementary Medicine Research Center, Mazandaran University of Medical Sciences, Sari, Iran.

Email: spsmazums@gmail.com

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1- INTRODUCTION

Steven-Johansson syndrome was first introduced as a special condition and epidermal eruption in 1922 (1). This syndrome is among the potentially fatal disorders which is associated with symptoms such as fever, extensive blisters in muscle tissues, vomiting and atypical signs like oral lesions and involvement of mucous membranes (2, 3). The disease is often accompanied with serious complications in organs such as liver, lung and kidneys that in the absence of timely diagnosis, treatment will be difficult and chance of recovery is reduced (4). The annual incidence of this syndrome has been reported to be 1 to 7 cases per one million people worldwide (5).

Infections are the most important factor causing this syndrome in children (6, 7). There are also reports of the effects of certain drugs (such as sulfonamides and barbiturates) on the occurrence of this syndrome in children (1). In total, more than 100 types of drugs causing this complication in adults and children are recognized (4).

Diagnosis of this syndrome is based on clinical signs on the one hand and also on histological features on the other hand. Initially, typical clinical symptoms include dark spots and positive "nikolsky" sign which is characterized by applying mechanical pressure on the induced skin and detachment of the epidermis and blisters within a few minutes to a few hours (7).

The treatment of Stevens-Johnson syndrome is based on two grounds: 1) eliminating the underlying factors; and 2) supportive therapy often with high doses of intravenous immunoglobulin (IVIG) and Plasmapheresis (8). However, the effectiveness of these treatments has been controversial (9). This article presents a case of Stevens-Johnson syndrome with idiopathic causes.

2- CASE REPORT AND RESULTS

The patient was a six-year-old, 154 cm tall and 18 Kg young boy who is the 1/1 child from non-related and healthy parents. He has been born by cesarean delivery and termed. The patient has had no problems in his fetal ultrasound. The subject was taken to the emergency of Bu-Ali hospital Sari (Mazandaran province, North of Iran) by his parents with complaining of fever and skin lesions since last two days. According to parents, the patient has been complaining of severe pain in the anus area which has exacerbated during disposal action. At the same time, skin lesions and blisters without itching have appeared around ears that gradually had spread to the face, trunk and limbs. He had painful aphthous ulcers in his mouth. The patient also had a fever and using antipyretic (acetaminophen) didn't have much impact on reducing his fever. The history of hospitalization and any underlying disease was not reported by parents. Patient`s vaccination has been completely done based on the country`s routine. No history of drug use was reported by the patient and his parents. The patient also had no history of drugs and food allergy. According to the child's parents, two days prior to the appearance of symptoms, they have used insecticides at home (manufactured by Tar-o-Mar company; containing permethrin, trans-alletrine, piperonyl butoxide, tetramethrin, sodium nitrite, sorbitan monooleate and water solvent).

The results of primary examinations of patients were as follows: BP: 100/60 mmHg; PR: 100 p/min; RR: 20 r/min; T: 39.2^{o}c (axillary); O_2 Sat: 99%. The subject was critically ill. Maculopapular and vesiculobullous lesions on the erythematous environment (some of them being crusted) were observed throughout the entire body. Some lesions also had necrotic center. Vesiculobullous lesions were seen in the external auditory canal,

but eardrum was normal. Also conjunctivitis was observed in the eye. Nasal mucosa wasn't pale and aphthous ulcers and vesicles were seen in oral cavity mucosa and patient`s palate. Gums were infamed (gingivitis) and there was no lymphadenopathy. Additional voices weren't heard on auscultation of the lungs and heart. The abdomen was soft and there was no hepatosplenomegaly. Erythema, vesicular lesions and white curds were observed in the anus area. "Nikolsky" sign was also positive. Ophthalmology and dermatology as well as CBC-diff, ESR, B/C, LFT, BUN, Cr, UA, Na, K and culture of skin secretions were asked for the patient.

Patient's test results were as follows: WBC: 10800/µL; Neut: 77%; EOS: 3%; Lymph: 18%; Hgb: 12.5 g/dL; Plt: 204000/µL; ESR: 32 mm/hr; Urea: 25 mg/dL; Cr: 0.6 mg/dL; AST: 32 U/L; Alt: 20 U/L; K: 4.2 mEq/L; Na: 138 mEq/L; UA: Normal; Wound discharge culture: E-coli>105; B/C: No growth.

At first, the differential diagnosis of Toxic epidermal necrolysis (TEN) and Staphylococcal scalded skin syndrome (SSSS) were advised for the patient. According to the patient's clinical symptoms (skin erythema distribution in the body) the existence of TEN syndrome, was rejected. Also, according to the lab results on the lack of growth of any bacteria in the patient's blood and urine, SSSS syndrome was rejected, too. Therefore, the patient was diagnosed with Stevens-Johnson syndrome was hospitalized in the isolation room under the supervision of physicians with expertise in pediatric infectious.

3- RESULTS

3-1. Therapeutic strategies and outcomes

For patients, IVIG (the first three days of treatment at a dose of 35 grams, then for two days at a dose of 15 grams and 8 grams per day for the next three days) vancomycin ampoule (360 mg intravenously injected every six hours), 9 mg of acetaminophen syrup, Al-Mg syrup in the amount of 3 cc and daily usage of betamethasone eye drops were prescribed. Daily compression of Lesions by 1:8000 Potassium permanganate following by applying of 2% mupirocin ointment on the lesions was done.

A combinatorial ointment containing betamethasone and vaseline was prescribed for the patient's lips. Due to wheezing in the lungs auscultation on the seventh day of hospitalization, using of ventolin nebulizer was started. Fever has subsided on the eighth day of hospitalization, so IVIG and vancomycin were discontinued and 250 mg cefuroxime tablets was started every twelve hours. Almost all lesions were dried on the ninth day of hospitalization. On the eleventh day, compression bypotassium permanganate solution was stopped. On the fourteenth day of hospitalization, the patient was discharged with the medication orders of using zinc sulfate syrup and saline nasal spray.

3- DISCUSSION

Taking certain medications is the most important cause of Stevens-Johnson syndrome (1), including sulfonamides, anti-seizures and anti-gout drugs (10). In the previous reports by Parveen (report of the effect of Lamotrigine), Duarte et al. (report of the effects of Carbamazepine), Powers et al. (report of the effect of Azithromycin), and Storim and colleagues (report of the effect of Unithiol) the role of drugs in causing Stevens-Johnson syndrome has been mentioned (11-14). However, report of the incidence of this syndrome is very rare without the usual reasons. In the present report, the subject has rejected any precedent drug use and the tests and the reports haven`t shown any

infection. However, genetic factors as one of the very rare causes of this syndrome have been emphasized in some studies (15). In this report, the patient had no pedigree of this disease in his family. This disease occurred after the use of insecticides at home. According to available databases so far, no link between insecticides and this syndrome have been reported. Treatment of Stevens-Johnson syndrome is based on receiving IVIG and infection and pain control are done by using broad-ranges of antibiotics and narcotic analgesics respectively (9). The reported subject was improved and discharged from the hospital after 14 days of receiving the above-mentioned medications.

4. CONCLUSION

The results showed that distraction technique had a good effect on the intensity of pain in children. Given the need for pain control and its effects on the course of treatment, further studies are needed to be done.

5- ABBREVIATION

BP: Blood Pressure,
PR: Pulse Rate,
RR: Respiratory Rate,
T: Temperature,
O_2 Sat: Oxygen Saturation,
CBC-diff: Complete Blood Count with Differential,
ESR: Erythrocyte Sedimentation Rate,
B/C: Blood Culture,
LFT: Liver Function Tests,
BUN: Blood Urea Nitrogen,
Cr: Creatinine,
UA: Urine Analysis,
K: Potassium,
N: Nitrogen,
Na: Sodium,
WBC: White Blood Cells,
Neut: Neutrophil,
EOS: Eosinophils,
Hgb: Hemoglobin,
Plt: Platelets,
AST: Aspartate Aminotransferase,
ALT: Alanine Aminotransferase.

6- CONFLICT OF INTEREST: None.

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