Early Treatment of Steven Johnson Syndrome Case in 2-Year-Old Boys: Case Report

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ABSTRACT
Introduction: Steven Johnson syndrome is a hypersensitivity reaction with rare, acute and life-threatening symptoms of skin and mucosal epidermolysis with 1 to 6 cases per 1 million cases annually. Case: A 2-year-old boy with complaints of vesicles on the back and abdomen that itch and sting accompanied by sores on the lips and genitals since 3 days ago. Patient's history of taking Amoxicillin, Guainefenesin, CTM, and Paracetamol. The patient had experienced the same complaint twice after being given the same medicine. Discussion: boys under the age of 3 years are very rarely found SSJ because SSJ is a slow type of hypersensitivity. Although rare in pediatric cases SSJ is found to have a higher mortality rate. Conclusion: Steven Johnson syndrome is a potentially life-threatening skin case. Cases of SSJ in children under 3 years of age are very rare because they are rarely exposed to drugs and are more often found in women.

1. Introduction

Access to getting medicines is now very easy. Various kinds of drugs in circulation have good and bad effects, one of the bad effects of drugs is causing allergic reactions such as Steven Johnson Syndrome (SSJ). Steven Johnson syndrome or Erythema Multiforme Majus, is a very rare, acute and potentially life-threatening skin and mucosal hypersensitivity reaction (Unnisa et al., 2021). Symptoms of SJS include extensive necrosis and peeling of the epidermis of the skin, orifice mucosa, and eyes caused by a type 3 hypersensitivity reaction or immune complex reaction (Goldsmith et al., 2012). The incidence rate of SJS is around 1 to 6 cases per 1 million in a year with a mortality rate ranging from 5% to 12% and the risk of increasing the mortality rate is found in patients aged more than 40 years, pulse more than 120 in a minute, history of cancer or malignancy, peeling of more than 10% of the body area, serum urea level >10mM, serum bicarbonate level >20mM, and serum glucose level >14mM. Mortality based on SCORTEN scoring varies depending on the number of points which can range from 3.2% - 90% (Goldsmith et al., 2012). Steven Johnson syndrome is often found in adults, whereas it is very rare in children under 3 years of age (Arwin et al., 2008). Hypersensitivity in SJS is often related to the use of drugs such as antibiotics, anti-seizure drugs, NSAIDS, and oxide inhibitors. Therefore, early recognition, diagnosis and treatment are very important for patient mortality. The following is a case of SJS caused by amoxicillin in children under 3 years.

2. Method

Case Illustrations

A 2-year-old boy came to the emergency room at Tabanan Regional Hospital with complaints of a rash appearing on his back and stomach in the form of watery spots which then broke and widened accompanied by itching and burning since 3 days ago. The patient complained of sores on his lips and a feeling of thickening making it difficult to eat accompanied by a fever of 37.8^{the}C, about 4 days ago the patient experienced fever and sore throat. The patient consumed medicines given by a general practitioner, namely Amoxicillin syrup, a mixture of Guainefenesin and CTM, and Paracetamol syrup. The patient's previous medical history has experienced the same condition twice after being given the same medication and this is the third time the patient has experienced a similar complaint. Dermatological examination of the thoracolumbar region multiple erythematous macules, round shape, well-defined, with a diameter of 3cm to 5cm, lenticular to nummular in size. Labia region, visible erosion, geographic shape, well-defined, lenticular size, wound surface covered with brownish crust. SCORTEN

scoring for patients was 0-1 with a mortality of 3.2%. The patient was diagnosed with SJS. The treatment given is liquid D5 ¼ NS 15tpm, Methylprednisolone 2x10mg, Amikacin Sulfate 2x135mg injection, Paracetamol 3x200mg orally and topical treatment is given Triamcinolone oral base 2x1, NaCl compress on the lips and body area, Triamcinolone oral base on the lip area after 10-15 minutes of NaCl compress.

Figure 1. On the left there is a lesion in the form of erosion spread in the region upper and lower lips. The right has multiple erythematous macular lesions in the region thoracolumbar

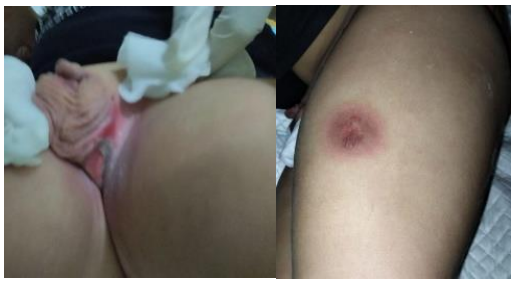

Figure 2. On the left there is a diffuse erythematous macular lesion accompanied by erosion in the region legs. Right There is an erythematous macular lesion accompanied by brownish crusting in the region left thigh

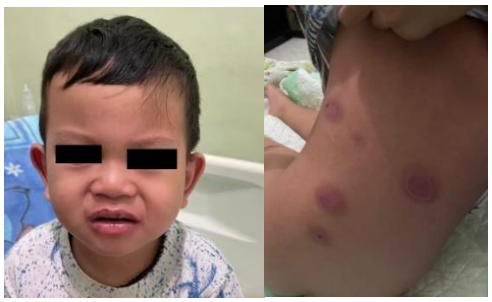

Figure 3. Left, lesion on the lips after day 3 of treatment. Right, lesion on the back after day 3 of treatment

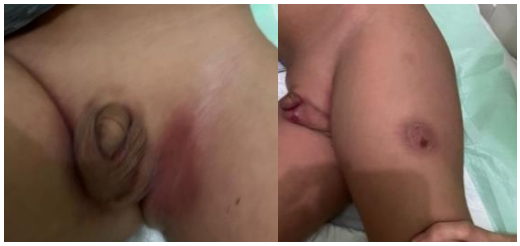

Figure 4. Right of the lesion between the thighs after the 3rd day of treatment. Right of the lesion on the left thigh after the 3rd day of treatment

Follow On the 3rd day, the lesion seemed to dry up and the redness began to fade, the patient also complained that it was no longer itchy and sore anymore and on the 5th day the patient was allowed to be outpatient.

3. Result and Discussion

Steven Johnson Syndrome (SSJ) is a life-threatening, acute condition, which generally affects the skin and mucous membranes, characterized by extensive necrosis and peeling of the epidermis of less than 10% of the

body surface area (LPB) due to a drug reaction or infection (Goldsmith et al., 2012; Unnisa et al, 2021)

The incidence of SJS is very rare, ranging from 1 to 6 per 1 million cases per year. SJS can affect people of all ages, but the risk will increase after the age of 40 years and it is very rare to find cases aged 0 to 3 years, women are more at risk, a history of immunodeficiency, a history of cancer and vascular collagen can also increase the risk.2 In the case of children with TEN or ages ranging from 0 to 5 years have the highest mortality of 7.5% to 23%.6 Mortality in SJS cases varies from 5% to 12%. SCORTEN is a prognosis scoring instrument that also measures mortality rates that is currently used (Goldsmith et al, 2012; Liotti et al., 2019).

Although the pathophysiology of SJS is currently still unclear, drug use is one of the most common etiologies apart from infections such as HIV, Herpes, systemic diseases, and genetics. Several classes of drugs are categorized as High Risk in causing the SSJ reaction to Toxic Epidermal Necrolysis (TEN), namely antibiotics, anticonvulsants, NSAIDs, Allopurinol, Oxicam, Lamotrigine, and Nevirapine.24

In children, it is very rare to find cases of SJS because SJS is a type 4 hypersensitivity reaction that requires repeated sensitization and exposure to finally erupt on the mucosa and skin (Goldsmith et al, 2012).

Although the pathogenesis of SJS is still unclear, several studies suggest a cell-mediated cytotoxic reaction against keratinocytes that causes massive apoptosis.

Clinically, in patients usually in the initial phase lesions will appear around the oral mucosa and eyes which are often ignored by the patient and family. Patients should seek treatment as early as possible to avoid severe SJS that can turn into TEN (Unnisa et al, 2021; Zankar et al., 2018).

In this case, there was a 2-year-old male patient who was diagnosed with overlapping SSJ-TEN with an expansion of 10%-30% LPB after having a history of taking the antibiotic Amoxicillin and had been treated for 4 days and experienced significant improvement.

4. Conclusion

Steven Johnson syndrome is a rare and potentially life-threatening skin condition. In this case, a case of SJS was found in a 2 year old boy, which is a very rare age for SJS reactions to occur. Without a history of immunodeficiency disease or cancer that could increase the risk of developing an SJS reaction, the risk should be very low. All cases of epidermolysis on the skin and mucosa with a suspected history of drug consumption need to be considered SJS or TEN as a diagnosis by ruling out risk factors according to theory and appropriate treatment can prevent morbidity and mortality in patients.

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