Case report

Valproate and Pediatric Stevens-Johnson Syndrome: A Critical Alert

ABSTRACT

This case highlights a three-year-old boy who presented to the emergency department with a fever, facial redness, and runny nose. With a previous history of trauma due to falls, it was found that the patient was on valproate. After consultation with dermatology, Steven Johnson syndrome was given as a diagnosis. The drug was discontinued and symptomatic treatment was initiated.

Keywords: Steven Johnson Syndrome, Valproate, Neurology.

1. INTRODUCTION

Stevens-Johnson Syndrome (SJS) is a rare but serious disorder affecting skin and mucous membranes. Typically triggered by a reaction to medication or an infection, SJS is characterized by a painful rash that spreads and blisters, leading to the top layer of skin shedding and falling off. This condition requires immediate medical attention, as it can lead to severe complications, including infections, dehydration, and organ damage. Early diagnosis and treatment are crucial for improving outcomes and preventing lasting damage [1].

Stevens-Johnson Syndrome often begins with flu-like symptoms such as fever, sore throat, and fatigue, followed by a red or purplish rash that spreads and forms blisters. These blisters can cause the skin to peel and affect the mucous membranes, including those in the mouth, eyes, and genitals. In severe cases, it can progress to Toxic Epidermal Necrolysis (TEN), a more severe form of SJS where more than 30% of the body surface area is affected. Immediate discontinuation of the offending drug and supportive care in a hospital setting, often in an intensive care unit or burn unit, are essential for management [2].

2. CASE REPORT

A three-year-old boy pediatric patient comes to the emergency department complaining of fever, facial redness, and runny nose. After introducing valproate due to a previous history of seizures and a history of jerking both limbs the patient developed facial redness, which involved the entire body on day four in the hospital. The patient is a known case of traumatic brain injury fall from the fifth floor in September 2022—developed seizures after acquired brain injury fall. There is no significant family history, personal history, or travel history. Current medication includes Levetiracetam, Valproate, Citicoline, and piracetam. A general examination pulse was 76 per minute, no available blood pressure, respiratory rate of 25 per minute, temperature was 36.2°C, weight was 18.8 kg, and appearance was distressed. On physical examination, there were no complaints of chest pain, palpitations, or cough, the abdomen was soft, non-tender, with no abdominal distension, alert oriented to please person time, please person time, quadriparesis, Spasticity with reduced speech output, Redness over the body.

Diagnostics were ordered EEG which revealed electrographic seizures, C-reactive protein (CRP): 6.3 mg/L (Reference range is Less than 5 mg/L), and Complete blood count revealed low platelet level 59 - 10^3 /uL (normal reference level is 200-490 10^3 /uL), and WBC count low 3.7 10^3 /uL (normal reference level is 5-15 10^3 /uL). Aspartate aminotransferase (AST) 139 which is high (Reference range is up to 50), and Alanine aminotransferase (ALT) 116 which is high (Reference range is up to 50). Kidney function test showed a low sodium level which was 131.5 (Reference range is 136-145 mmol/L), and Albumin was low at 3.64 (Reference range is 3.8-5.4 mmol/L).

After a Consultation with a Dermatologist Steven Johnson Syndrome was given as diagnosis due to the intake of valproate.

3. DISCUSSION

Stevens-Johnson Syndrome (SJS) is a rare life-threatening condition characterized by severe mucocutaneous epidermal necrolysis and detachment of the epidermis. The condition centers around a delayed-type hypersensitivity reaction with a complex etiology stemming from a variety of causes [3].

Early SJS signs are challenging to diagnose, often resulting in delayed treatment. Initial symptoms can appear 1-3 weeks after drug exposure, with mucosal lesions leading to more severe skin reactions. Diagnosis is confirmed through a skin biopsy. The acute phase lasts 8-12 days, followed by re-epithelialization. Treatment includes immediate drug cessation, supportive care, and possible burn unit transfer. Mortality rates are under 10% for SJS and 30-50% for TEN, with co-infection and respiratory failure as common causes of death. Long-term complications include ocular issues and adhesions in gastrointestinal and urogenital tracts [4].

SJS pathophysiology involves widespread keratinocyte apoptosis mediated by T lymphocytes and natural killer (NK) cells. Granulysin is identified as a key cytotoxic molecule in blister fluid correlating with disease severity. The SCORTEN scale predicts mortality, ranging from 3.2% with low scores to 90% with high scores [3].

A systematic review of Stevens-Johnson Syndrome (SJS) and Toxic Epidermal Necrolysis (TEN) associated with Valproic acid included case studies primarily from India, Turkey, China, and Italy to find the prevalence for SJS/TEN with different antiepileptics. They concluded that Phenytoin, lamotrigine, and carbamazepine are common antiepileptics linked to these conditions, while valproic acid (VPA) is less frequently associated. The defect in epoxide hydrolases prevents the excretion of toxic metabolites, leading to hypersensitivity reactions and skin lesions [4].

4. CONCLUSION

In this case, a three-year-old boy presented with Stevens-Johnson Syndrome (SJS) after the administration of valproate for seizure management following a traumatic brain injury. SJS, while rare, is a severe condition that necessitates prompt recognition and treatment. The initial presentation with fever, facial redness, and a runny nose, combined with a significant history of seizures and valproate usage, raised suspicion for SJS, confirmed by dermatology consultation.

Management of SJS involves immediate discontinuation of the offending drug, in this case, valproate, and initiation of supportive care to manage symptoms and prevent complications. The complexity of SJS's pathophysiology, involving immune-mediated keratinocyte apoptosis, underscores the importance of early diagnosis and intervention to improve patient outcomes. Despite its rarity, healthcare providers must remain vigilant for SJS signs, particularly in patients receiving medications known to trigger this syndrome.

This case underscores the importance of considering SJS in pediatric patients with skin and mucous membrane involvement following drug administration. Early dermatological consultation and multidisciplinary care are crucial for managing this life-threatening condition effectively. Further research and awareness are essential to understand better the risk factors and mechanisms underlying drug-induced SJS to improve prevention and treatment strategies.

CONSENT

Consent was taken from the patient for the publication of the case report.

ETHICAL APPROVAL

Not Applicable

REFERENCES

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