Case Report of Atypical Steven Johnson Syndrome in a Paediatric Patient

Isabella Tso [1], Carina Lam [1]

[1] School of Medicine, University College Cork, Cork, Ireland

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Abstract

Introduction: Steven-Johnson Syndrome (SJS) is a rare mucocutaneous condition with variable severity. While medications are the most common triggers, aetiologies differ between paediatric and adult populations. This case report describes an atypical presentation of SJS in a 10-year-old male in Cork, Ireland, and explores differences in triggers across age groups.

Methods: Both the patient and the guardian provided informed consent to share this information for educational purposes.

Results: The patient presented with an atypical, severe form of SJS, including progressive lip oedema that was minimally responsive to prednisolone, alongside evolving black haemorrhagic bullae. Blood cultures identified Mycoplasma pneumoniae. With a Naranjo Adverse Drug Reaction Probability Scale score of 2, a drug-induced cause was considered unlikely, particularly in the context of a concurrent infection. Further investigations showed no underlying immunological or chronic conditions.

Discussion: In adults, SJS is most commonly triggered by medications such as non-steroidal anti-inflammatory agents (NSAIDS), anti-epileptics, and antibiotics. In contrast, paediatric SJS is more often associated with infectious agents, with Mycoplasma pneumoniae being the most frequent bacterial cause. Recognizing this distinction is vital for diagnosis, prevention, and management strategies.

Conclusion: Although SJS is rare, it can have profound impacts on patients. Awareness of the differences in triggers between children and adults is crucial for effective prevention and management, and helps guide both clinicians and families in decision-making.

Introduction

Steven-Johnson Syndrome (SJS) is a rare type IV hypersensitivity immune response affecting the skin and mucous membranes (1). Despite its low prevalence, SJS disproportionately affects children and young adults, with an incidence of 1.2-6 cases per million per year (2).

Initial symptoms typically include fever, malaise, sore throat, nausea, vomiting, arthralgia, and myalgia (1,3). These are often followed by conjunctivitis and mucosal bullae, particularly involving the oral cavity, nares, and pharynx (3).

The presence of open lesions increases the risk of secondary infection. Morbidity is significant, with mortality rates reported up to 5% (3). Therefore, early recognition and intervention are essential.

Although the precise pathogenesis remains unclear, effective treatment relies on identifying the underlying trigger (4). Medications are most frequently implicated, making prompt discontinuation critical (4). However, triggers vary by population.

In paediatric patients, infections are more common causes, with Mycoplasma pneumoniae (M. pneumoniae) being the most frequently identified bacterial trigger, despite extrapulmonary manifestations occurring in only 6.8% of cases (5).

This case report describes a 10-year-old male presenting with atypical SJS at Cork University Hospital (CUH), suspected to be triggered by M.pneumoniae. It also highlights the differences in SJS triggers between adult and paediatric populations.

Case Background

A 10-year old male who presented to CUH with atypical SJS.

Presenting Complaint

R.M., a 10-year-old male, presented to the CUH emergency department (ED) with a two-day history of worsening erythematous, swollen lips with black haemorrhagic bullae, minor ulceration, bilateral conjunctivitis, and fever, on a background of a one-week history of worsening odynophagia.

History of Presenting Complaint

Two weeks prior, R.M. developed a non-productive cough, thought to be a lower respiratory tract infection (LRTI), and was prescribed a 5-day course of co-amoxiclav (amoxicillin–clavulanate) 312 mg three times daily by his general practitioner.

Within 24 hours of starting antibiotics, he developed lip swelling without associated respiratory or other anaphylactic features. He subsequently attended SouthDoc, where antibiotics were discontinued and subsequently commenced on 15 mg oral prednisolone for the oedema, which mildly improved initially.

Over the following 48 hours, his symptoms progressed, with development of haemorrhagic bullae and dysphagia, prompting presentation to the ED.

Past Medical History

His father denied any known drug allergies, regularly prescribed medications, or medical conditions.

For previous infections, R.M. has received co-amoxiclav and other antibiotics with no reactions or concerns. R.M. is up to date on all vaccines, and there is no relevant family history of autoimmune conditions such as Sjögren syndrome.

Social History

R.M. is selective with his food intake, preferring a brown diet, but continues to track appropriately for his height and weight centiles.

Clinical Findings

On exam, R.M. was vitally stable and remained afebrile. On visual inspection, R.M. had bilateral pruritic conjunctiva, an erythematous throat, severely edematous lips with open blisters and hemorrhagic bullae, limiting his ability to open his mouth. Further examination yielded no other dermatological findings on the peripheries.

Investigations

Initial blood tests showed an elevated white cell count, lymphocytosis, thrombocytopenia, and raised inflammatory markers. Immunological investigations were unremarkable, with a normal CD4 count. Viral swabs, including tests for herpes simplex virus (HSV), were negative. Blood cultures were positive for M. pneumoniae, prompting a 5-day course of oral azithromycin treatment. Skin biopsy was not performed due to the atypical presentation isolated to the lips. On the Naranjo Adverse Drug Reaction Probability Scale, R.M. scored 2 (Table 1), suggesting a possible but unlikely drug-related cause

Treatment

R.M. was admitted for 10 days. During his admission, he received:
5 days of prednisolone (1 mg/kg) for lip oedema
1 day of oral aciclovir (discontinued following negative HSV results)
5 days of azithromycin for M. pneumoniae

Benzydamine (Difflam) spray was used for symptomatic relief. NSAIDs were avoided due to their potential to trigger or worsen SJS.

Table 1 Naranjo Adverse Drug Reaction Probability Scale Parameters and Corresponding RM Score Calculation

Question	Answer (Yes, No, Unknown)	Score
Are there previous conclusive reports on this reaction?	No	0
Did adverse events appear after the suspected drug was given?	Yes	+2
Did the adverse reaction improve when the drug was discontinued or a specific antagonist was given?	Yes	+1
Did the adverse reaction appear when the drug was readministered?	Unknown	0
Are there alternative causes that could have caused the reaction?	Yes	-1
Did the reaction reappear when a placebo was given?	Unknown	0
Was the drug detected in any body fluid in toxic concentrations?	No	0
Did the patient have a similar reaction to the same or similar drugs in any previous exposure?	No	0
Was the adverse event confirmed by any objective evidence?	No	0
Total Score	2	

Discussion

Over 100 medications have been linked to the development of SJS and toxic epidermal necrolysis (TEN). Common culprits include sulfonamides, anticonvulsants (such as phenytoin, phenobarbital, and carbamazepine), NSAIDs, and allopurinol (6). Genetic factors, particularly specific human leukocyte antigen (HLA) allotypes, have been associated with an increased risk of developing SJS (Table 2), especially when these individuals are exposed to aromatic anticonvulsants and allopurinol. Select viruses can also trigger SJS, but the pathogenesis remains poorly understood. There is evidence suggesting that these viruses can trigger the condition independently of drug exposure. It is hypothesized that viral infections induce a severe immune response, leading to the characteristic symptoms of SJS (7). Additionally, viruses may act as a catalyst, modifying the immune system's response to other triggers and potentially increase SJS risk (7). The dual role highlights the complexity of SJS pathogenesis and the importance of understanding how viral infections influence immune responses.

Identified viruses associated with SJS complications include influenza virus, varicella virus, COVID-19, and coxasackievirus (7). Recently, there have been reported cases of M.pneumoniae infections associated with extrapulmonary complications, including SJS. Many patients presented with polymorphic or vesiculobullous lesions. Other bacterial species, such as Staphylococcus aureus and Acinetobacter baumannii, are often isolated from SJS patients during hospitalizations (8,9). These are typically linked to secondary infections (e.g., skin or bloodstream infections) rather than acting as primary triggers of SJS (8,9). There are many causes of SJS, however, triggers differ between the adult and paediatric populations. Drug-induced SJS is more common in adults, whereas infection-triggered SJS is more frequent in children. In adults, most cases are associated with medications such as antibiotics (particularly beta-lactams, sulfonamides, and fluoroquinolones), anticonvulsants, allopurinol, and NSAIDs. These account for up to 68% of cases and are associated with higher morbidity and mortality (10).

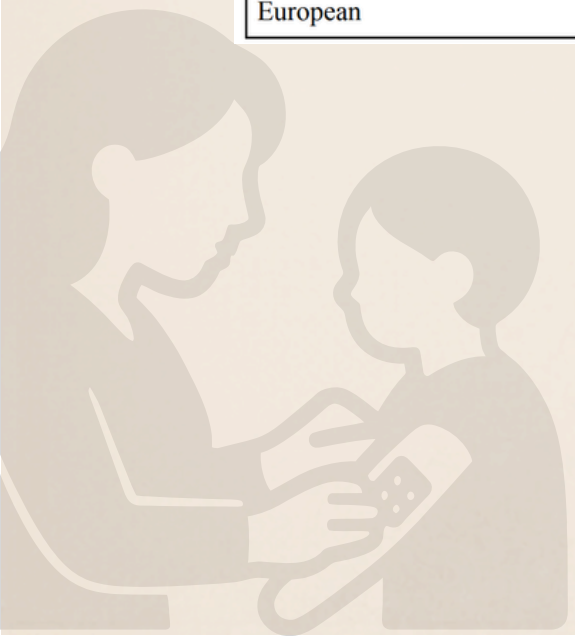
In contrast, approximately 43-53% of SJS pediatric cases are medication-related. Infections, particularly M.pneumoniae, are a significant trigger in the paediatric cohort. Infectious etiologies are predominant in children, with M.pneumoniae causing 19-22% of cases while HSV accounts for approximately 9% (5,11). In summary, adults are more likely to develop SJS due to drug exposure, whereas children are more likely to develop SJS secondary to infections, particularly from M.pneumoniae, as demonstrated in this case.

Conclusion

This case describes a 10-year-old male who developed SJS with prominent mucositis following a M.pneumoniae infection. His presentation, characterised by severe lip oedema, haemorrhagic bullae, bilateral conjunctivitis, and odynophagia with minimal cutaneous involvement, represents an atypical but significant manifestation. The positive blood culture, negative viral studies, and low Naranjo score support an infection-triggered rather than drug-induced cause, despite recent antibiotic exposure. This case highlights an important age-related difference in SJS aetiology. While drug-induced SJS is more common in adults, infectious triggers—particularly M. pneumoniae—are more frequently seen in paediatric patients. Recognising this distinction is important, as it has direct implications for diagnosis, management, and future medication safety.

Table 2 Demographic and Corresponding HLA Allele Variant with the associated SJS triggering Drug Class (1,3)

Ethnicity	HLA allele	Drug
Han Chinese Thai Malaysian South Indian	HLA-B*1502	Aromatic anticonvulsants
Han Chinese	HLA-B*5801	Allopurinol
European	HLA-B*5701	Abacavir
European	HLA-A*3101	Carbamazepine



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