

An Atypical Trigger of Stevens-Johnson Syndrome with Severe Ocular Complications

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Abstract

Presentation

We present a rare food-related case of an 18-year-old male with widespread rash including the oral mucosa.

Diagnosis

Diagnosis was based upon the progression of mucocutaneous symptoms and ocular findings confirmed by ophthalmology, and the exclusion of alternative diagnoses through negative vasculitis and viral screens.

Treatment

Treated with intravenous corticosteroids, antibiotics, fluid resuscitation, dexamethasone eye drops, and underwent bilateral amniotic membrane transplantation for sight-threatening ocular involvement.

Discussion

This case highlights key learning points in the management of Stevens-Johnson Syndrome (SJS), including the importance of early specialist referral, cautious antibiotic use in suspected cases, and maintaining a high index of suspicion for atypical triggers such as food, all of which can significantly impact patient outcomes.

Introduction

SJS often starts with flu-like symptoms and rapidly progresses to blistering skin and mucosal erosions, with eye involvement in up to 90% of cases^{1,2}. A significant proportion of cases are associated with several medications including anticonvulsants, Penicillin-based antibiotics, as well as Sulphonamides and Allopurinol^{3,4}. In cases where epidermal detachment is less than 10% of the total body surface area it is categorised as SJS. This contrasts with Toxic Epidermal

Necrolysis (TEN), where the affected surface area is more than 30% of the body⁵. The pathogenesis of SJS is not fully understood, however it is thought to be related to a cytotoxic immunological response mediated by CD8+ T cells and an association of genetic factors such as HLA haplotypes⁶. Less commonly, SJS/TEN can be caused by pathogens, with bacterial infections being the most common pathogenic cause of SJS⁷.

Case Report

An 18-year-old male presented to A&E with a rash to his whole body including his limbs, trunk and mouth that developed over several hours. He denied any recent illness or medication use, however mentioned ingestion of a bottle of expired camel milk a day prior to symptom onset. Figures A and B demonstrate the extent and distribution of the mucocutaneous rash observed in this case.



Figure 1. (A) Diffuse symmetrical erythema and coalescing maculopapular lesions on palms bilaterally; **(B)** Extensive mucosal involvement, with haemorrhagic crusting and ulceration of the lips and erythematous, crusted lesions on the nose.

Initial blood results displayed a raised C-reactive protein (CRP) and the patient was managed as suspected SJS with intravenous corticosteroids, Co-amoxiclav and fluid resuscitation. We have calculated the SCORTEN score to be 0.

Following post-take ward round, dermatologist review was sought. This request came 6 hours post initial presentation. Vasculitis and viral screens were both negative, after which systemic steroid treatment was stopped. The following day, the patient complained of oral ulceration, painful penile erosions and purulent eye discharge and was reviewed by an ophthalmologist one week later.

Ophthalmologist review noted bilateral ocular pseudo-membranes, symblepharon and painful bleeding of the lips with specialist confirming clinical diagnosis of SJS. The severity of eye involvement was sight threatening, and bilateral amniotic membrane transplant was arranged with interim dexamethasone eye drops.

Eight days following admission the patient was reviewed in dermatology clinic, however no formal advice or clinic letter was received. The patient later developed onycholysis and was similarly reviewed in dermatology clinic a month after admission, with skin lesions having resolved by this point. The patient was discharged with dermatology outpatient follow up.

Discussion

The commonest causes of SJS are drugs (53-95%), infection (30%) and idiopathic (5-18%)⁸. Extensive screening tests did not yield a causative drug or infectious agent, with the only associated detail being ingestion of expired camel milk. Food causes are rare with only one report associated with ingestion of ginger and herbal plants⁹. Early recognition is key in managing SJS. Prompt corticosteroid therapy was initiated, a strategy known to curb disease progression with minimal adverse risk¹⁰.

Studies report the importance of early specialist review. In our case ophthalmology review took place one week after initial presentation, a delay which is likely to have contributed to symptom severity, with the patient ultimately needing bilateral amniotic membrane transplant.

In conclusion, the evidence is not robust, but it suggests possible association of SJS with expired camel milk. The major learning point of this case is the need and awareness of a low threshold for prompt specialist referral. This will lead to early diagnosis, treatment and prevent serious ophthalmological complications of SJS. It also provides evidence to a possible link of food triggers to SJS.

Declaration of Conflicts of Interest:

None declared.

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