Beyond the Rash: The Fatal Consequences of Lyell Syndrome

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Abstract

Lyell syndrome is a rare, unpredictable, severe, and potentially fatal disease. Diagnosis is primarily clinical, confirmed by pathological examination, and lacks specific biological abnormalities. Several drugs, including allopurinol, are implicated in its onset. Therapeutic management requires admission to an intensive care unit, ideally a specialized burns resuscitation unit, and is essentially symptomatic. Treatment focuses on analgesia, hydro electrolytic resuscitation, nutritional support, wound care, infection prevention, psychological support, and social reintegration.

Keywords: Lyell Syndrome, Necrosis, Outcomes, Necrosis

Introduction

Toxic epidermal necrolysis (TEN), also known as Lyell syndrome (LS), is a rare, unpredictable, severe, and potentially fatal form of toxidermia [1]. It is the most severe form of drug-induced skin reaction, with a mortality rate of up to 25% and long-term sequelae affecting 80% of survivors, primarily involving ocular, genital, cutaneous, and bronchial complications [2]. LS is characterized by extensive epidermal necrolysis affecting at least 30% of the skin surface, often associated with erosive mucosal damage. The most commonly implicated drugs include trimethoprim, sulfamethoxazole, allopurinol, anticonvulsants, and penicillins [3]. Management requires admission to an intensive care unit, preferably a burns resuscitation unit, and is predominantly symptomatic, focusing on pain control, fluid and electrolyte management, nutritional support, wound care, and infection prevention [4]. We report a case of Lyell syndrome secondary to oral allopurinol.

Case Report

A 59-year-old woman with a history of arterial hypertension for over ten years was referred to the Emergency Department for extensive skin detachment (Figure 1).

Initially, the patient had consulted for polyarthralgia, prompting laboratory investigations that revealed hyperuricemia. Consequently, she was prescribed allopurinol for a suspected gout attack. On the fifth day of treatment, she developed body aches and erythema (Figure 2),

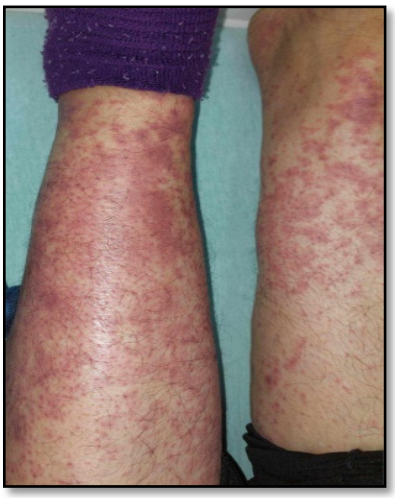

Figure 1: Extensive skin detachment

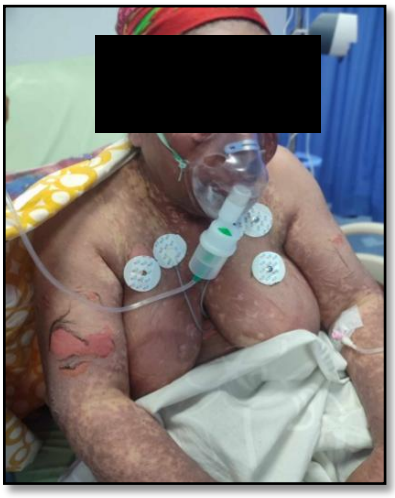

Figure 2: Pigmentation and skin eruptions

which progressed to diffuse pruritic skin eruptions, epidermal detachment, dysphagia, hypersalivation, and fever. She presented to the Emergency Department and was subsequently admitted.

Upon admission, the patient was in an altered state, tachycardic (110 beats/min), tachypneic (22 breaths/min), and had an oxygen saturation of 94%. Her blood pressure was 130/80 mmHg, and she was febrile at 38.9°C. Dermatological examination revealed extensive epidermal detachment resembling wet linen over a mildly erythematous base, covering more than 70% of the body surface. The Nikolsky sign was positive, and there were erosions of the ocular, buccal, genital, and anal mucosa (Figures 1, 2, 3, and 4).

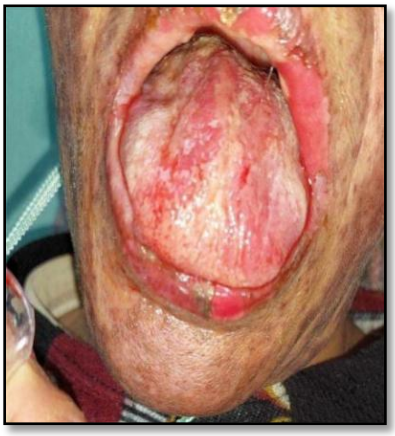

Figure 3: Buccal eruptions

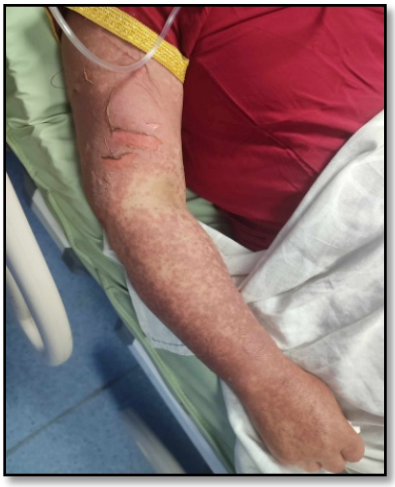

Figure 4: Extensive skin detachment

Laboratory investigations showed an inflammatory response with a C-reactive protein (CRP) level of 190 mg/L. Blood glucose was 10.9 mmol/L, urea 19 mmol/L, and bicarbonate 26 mmol/L. The case was reported to the regional pharmacovigilance center. Blood cultures were positive for Staphylococcus aureus, while urine cytobacteriological examination (UCE) was negative. The patient received appropriate antibiotic therapy. Despite ten days of intensive

resuscitation, daily dermatological care with silver sulfadiazine, Vaseline ointment, and ocular and oral care, the patient's condition deteriorated, and she ultimately succumbed to the disease.

Discussion

Allopurinol is a uricostatic agent that inhibits xanthine oxidase, an enzyme responsible for uric acid biosynthesis. It is metabolized to oxypurinol, which also inhibits xanthine oxidase, contributing to its therapeutic effect [5]. Allopurinol is one of the most frequently implicated drugs in Lyell syndrome. In our patient, its causative role was supported by the chronological sequence of events and the absence of other drug exposure in the two months preceding symptom onset.

Lyell syndrome is a rare disease, and its diagnosis is primarily clinical, confirmed by pathological examination, with no specific biological markers [6]. Histopathological analysis is essential to confirm the diagnosis, which has significant medical and legal implications. Due to the disease's rarity, it is often underrecognized, leading to delayed diagnosis and management [7]. Our patient met the widely accepted diagnostic criteria and had a SCORTEN score of 3, indicating a 35% risk of mortality.

Visceral involvement is a severe prognostic factor, manifesting as congestive erythema, erosions, and ulcerations of the digestive tract mucosa, pseudomembranous colitis, or respiratory mucosal involvement with acute respiratory distress syndrome, potentially complicated by infections. Fatal outcomes are typically associated with multiorgan failure, including pulmonary, cardiac, hepatic, and gastrointestinal failure. Early and multidisciplinary management can significantly improve outcomes [6].

For survivors, long-term sequelae are common and can be aesthetic, functional, psychological, and social. These sequelae affect approximately half of all patients [6,7] and may include cutaneous (dyschromia and superinfection-related pigmentary changes), ophthalmological (xerophthalmia, photophobia, keratitis, and potential visual impairment), genital (dyspareunia, synechiae, dryness, persistent erosions), sensory (taste disturbances, sweating abnormalities, nail disorders), and psychological (drug phobia) complications. Our patient experienced dysphagia, eye pain, and photophobia prior to her deterioration.

Conclusion:

Lyell syndrome remains a rare but severe condition with high morbidity and mortality rates. Early management in an intensive care unit, ideally a specialized burns resuscitation unit, is crucial. Treatment is primarily symptomatic, focusing on analgesia, fluid and electrolyte resuscitation, nutritional support, wound care, infection prevention, psychological support, and social reintegration.

Conflicts of interest: The authors have no conflicts of interest to declare.

Right to Privacy and Informed Consent: The authors have obtained the written informed

consent of the patients or subjects mentioned in the article.

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