

Case Report

Lyell's syndrome: a rare case report

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ABSTRACT

Lyell's syndrome or toxic epidermal necrolysis (TEN) is an epidermal detachment of more than 30% of total body surface area, most commonly due an idiosyncratic immune-allergic reaction to a drug. It causes an acute necrosis of the epidermis, with a clinical picture resembling extensive burns. There is also associated mucosal damage and can occasionally develop into a multiorgan having a poor prognosis. Here we report a case of toxic epidermal necrolysis, due to allopurinol intake and its management as in a large burns and stressing the importance of early treatment in an intensive care unit, with high vigil on fluid and electrolyte balance, prevention and treatment of infection with appropriate antibiotics and regular dressings.

Keywords: Toxic epidermal necrolysis, Emergency, Poor prognosis

INTRODUCTION

In 1956, a Scottish dermatologist Lyell first described this syndrome. He observed that in 4 patients with skin peeling due to epidermal necrolysis of toxic origin, presented with a clinical picture of burns, and that the causes for SJS and TEN were same.¹ Therefore, SJS and TEN were variants of a serious illness, differing only by the extent of cutaneous detachment (<10% of the total area for SJS, more than 30% for TEN). Between 10-30% was the transition form. Lyell's syndrome is a rare but serious adverse drug reaction with unpredictable occurrence leading to a destruction of the superficial layer of the skin and mucous membranes.^{2,3} Many drugs that may be responsible included allopurinol, sulfonamides, antiepileptic (carbamazepine, phenytoin, phenobarbital), non-steroidal anti-inflammatory drugs, etc.^{4,5} The management of this condition is mainly symptomatic. It similar to that of treating severe burns, with stopping of the suspect medication, fluid and electrolyte balance, prevention and appropriate treatment

of the infection and regular closed dressings. The prognosis is poor and is evaluated using a specific scale of severity of the disease, the SCORTEN.⁶

CASE REPORT

A 36 year old female presented to us from the dermatology department with entire skin showing epidermal peeling with blisters, similar to an extensive burn wound. She was diagnosed to have gout elsewhere for which she was started on allopurinol tablets since 5 days. Initially, she developed an erythematous rash which became bullous and started peeling and blistering after 2 days, when she presented to us (Figures 1-3). She had large cutaneous ulcers over the entire body which were of mixed depth exposing the deep dermal layers in a few areas. There was serous fluid discharging from these ulcers which were also tender. She was not able to walk or do her daily activities. There was edema of the body with involvement of the oral mucosa as well. All her investigations were normal. She was treated like a burn

victim with high protein diet, plenty of oral fluids, intravenous fluid replacement, parenteral antibiotics and regular debridements under anaesthesia and closed paraffin and compression dressings. She was given appropriate and aggressive physiotherapy and mobilization. The wounds were healing well and after about 3 weeks, all wounds healed well with minimal scarring (Figures 4-9).



Figure 1: Patient with cutaneous ulcers in the trunk and upper limbs.



Figure 2: Entire anterior body and lower limbs showing ulcers.



Figure 3: Back and gluteal regions showing involvement.



Figure 4: Healed trunk region.



Figure 5: Healed right upper limb.



Figure 6: Healed left lower limb.



Figure 7: Healed back and gluteal regions.



Figure 8: Healed anterior aspect of lower limb.



Figure 9: Healed gluteal region and posterior aspect of lower limbs.

DISCUSSION

Lyell's syndrome is a rare syndrome causing a severe mucocutaneous exfoliation and occurs in about 1 to 2 cases per million annually.⁷

It is a serious and grave drug allergy and is unpredictable. This condition destroys the most superficial part of the skin. The mucous membranes of the oral, ocular and urogenital region are also affected. The disease extends for a few days to a week. The severity depends on the extent of involvement of the skin and mucous membranes. The re-epithelialisation of the epidermis is rapid averaging 2 to 3 weeks.

Clinically skin lesions predominate in the face, neck and trunk, spreading rapidly to all layers with the sudden onset of large epidermal detachment exposing a red weeping dermis, which was the case in our patient.^{7,8} The development of the cutaneous symptoms was classical. The skin lesions were clean and not infected and there was complete healing after 15 days.

Drugs being the causative factor of this syndrome is about 60-70% of cases. The onset time is 1 to 4 weeks after the beginning of the drug taken with an average 2 weeks. The condition may be delayed for several days after stopping the drug if its half-life is long. When the treatment is taken for more than 2 months, the risk is minimal. The occurrence is independent of the dose and minor exposure is sufficient to trigger the condition.^{9,10}

According to data from the literature, almost all antibiotics can be indicted, with a particular frequency for penicillins, quinolones, vancomycin, macrolides and antituberculous drugs.^{9,12-15} Other drugs include allopurinol, carbamazepine, phenobarbital, phenytoin, and nonsteroidal anti-inflammatory drugs, pantoprazole and tramadol. Among newer drugs reported are nevirapine and lamotrigine and sertraline.

The incidence of this syndrome is rare with 0.4–1.4 cases/million/year, and the mortality rate ranges from 25% to 70%, depending on age, initial body surface area of epidermal detachment and comorbid factors.¹¹

In our patient, she was given allopurinol for gout after which she developed TEN.

Parenteral nutrition is often seen required in patients with oral lesions. We need to administer a high protein diet as this is a hypercatabolic state.

Most important is wound care done by saline wash and closed paraffin dressings in the operation theatre, till wound is healing well. Currently, there are no specific treatment that are established. The use of systemic corticosteroids debatable and various immunosuppressive therapies like cyclophosphamide and cyclosporine have been tried in few cases without much results. The use of

high doses of intravenous immunoglobulins have not been confirmed. The prognosis remains poor, but outcome depends on factors like age, the extent of the skin peeling, the delay in diagnosis and management, late discontinuation of the drug and infections which is a reason for a high mortality rate. It is evaluated using a specific scale of severity of the disease, the SCORTEN (Table 1).¹⁶ It consists of seven clinical and laboratory parameters that must be collected within 24 hours after admission.

The most significant effects are scarring with hyperpigmentation and corneal sequelae (pillowcases, white spots, which may require a corneal transplant).

A score lower or equal to 2 indicates a probability of survival about 90%, and when it is upper or equal to 4 the probability of survival is lower than 50%.

CONCLUSION

Lyell's syndrome is a rare but grave condition, associated with high morbidity and mortality. The importance of early management of patients with TEN in an intensive care unit is clearly understood as in our case. It is also important to inform the patient and their family about the need to inform the treating physician regarding the condition prior to starting any form of medications. This will reduce the incidence of this syndrome and improve prognosis by early treatment.

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