Clinical Presentation

A 75-year-old man presents to the Emergency Department (ED) with 3 days history of developing skin bullae that are peeling off with minimal serous fluid and minimal bleedings. The patient had runny nose and myalgia for which he received a diclofenac sodium injection 24 hour before the onset of his skin lesions. (Figure 1 & 2)

Figure 1: The initial bullae coalesced, leading to extensive exfoliation of the epidermis.

Figure 2: Extensive bullae in upper extremity

Questions

1. What is the diagnosis?
2. What physical sign that can be elicited in this patient?
3. How would you treat this condition?

Answers to clinical quiz on page 119
Answers

1. Toxic Epidermal Necrolysis
2. Nikolsky's sign is positive in patients with Toxic Epidermal Necrolysis. This sign can be elicited by applying a pressure in a sliding motion to the lateral aspect of a bulla which causes its extension. This is due to the loss of normal cohesion between epidermal cells and the basal layer that might result in erosions.
3. Discontinuation of the offending agent, immediate fluid resuscitation and recently immunoglobulin is considered in the treatment of TEN which is found to halt the progression of lesions and shorten the course of the illness.

Discussion

Toxic epidermal Necrolysis (TEN) is characterized by skin bullae and diffuse exfoliation of cutaneous surface and involvement of the mucous membrane. It is mostly caused by drugs. More than 100 medications have been identified as culprit agents, including sulfa groups, anti-convulsants (phenytoin, carbamazepine and phenobarbital), NSAIDs, allopurinol and antibiotics (cephalosporins, fluoroquinolones, and penicillins). Immunohistological studies have shown a widespread dermal infiltrates in patients with Toxic Epidermal Necrolysis suggesting a cytotoxic cell-mediated reaction against keratinocytes. This cell-mediated immune response leads to keratinocyte death by apoptosis.

The most important step in the management of TEN is to identify the offending agent and discontinuing it and immediate fluid resuscitation. Other adjuvant treatments are complementary. These include: Plasmapheresis and immunosuppressive drugs. Recently literature has shown that early treatment of immunoglobulin with 1 g/kg/day for three days improves the outcome dramatically. It stops the development of new bullae, halts disease progression and shortens the course of the illness.

References