Stevens-Johnson Syndrome and Toxic Epidermal Necrolysis: Oncologic Considerations

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Stevens-Johnson syndrome (SJS) and toxic epidermal necrolysis (TEN) are advanced forms of erythema multiforme major (EMM), a condition that causes skin eruptions. In SJS and TEN, eruptions become confluent, affecting the epidermal layer of the skin and mucous membranes, and resemble a burn. The majority of cases result from a drug reaction, and many of these drugs are used to treat patients with cancer. SJS occurs in one to six cases per million, whereas TEN occurs in 0.4–1.2 cases per million (Ellis, Oster, Turiansky, & Blanchard, 2002). These diseases often are misclassified, which may be, in part, because very few physicians have ever seen a case of SJS or TEN (Patterson & Tripathi, 1999). In general, classifying the diseases correctly depends on the presence or absence of mucous membrane involvement, number of membranes involved if mucous membranes are affected, and percentage of body surface area involved.

SJS was named for two pediatricians who documented the condition in children in 1922 (Helvig & Mann, 1998). SJS and TEN are rare, but both represent severe manifestations of a hypersensitivity reaction, usually to a drug, which requires complex medical and nursing care. Early recognition and interventions can significantly alter the course of the disease and improve the outcome. In patients with cancer, these diseases should be treated as oncologic emergencies.

Stevens-Johnson syndrome (SJS) and toxic epidermal necrolysis (TEN) are severe skin toxicities that may occur in patients with cancer. They are caused by infection or a drug reaction and can result in sepsis, severe ocular complications, and even death. Skin lesions usually are preceded by prodromal flu-like symptoms. A rash with subsequent blistering and denudation follows. Diagnosis is made by skin biopsy, and classification is based on body surface area and visceral organ involvement. Because many of the drugs associated with SJS and TEN are used to treat cancer, early recognition and intervention are critical to achieving a favorable outcome. Interventions include stopping the suspected offending agent. The use of steroids is controversial. Healthcare professionals always should consider an early transfer to a burn unit for patients with a comorbid condition such as cancer because this action is associated with improved outcomes. Implications for oncology nurses and a case study are presented in this article.

Key Words: Stevens-Johnson syndrome; epidermal necrolysis, toxic

Etiology

Pediatric cases of SJS are related more often to infection, whereas adult cases typically are drug or malignancy related. In about 50% of cases, no identifiable cause is found (Parillo & Parillo, 2002). The drugs most commonly associated with these diseases are anticonvulsants, sulfia preparations, allopurinol, corticosteroids, and non-steroidal anti-inflammatory drugs (see Figure 1).

Symptoms usually appear within the first 14 days of the start of the offending drug therapy (Bankston, Deshotels, & Daughtry, 2001). Both conditions also are associated with bacterial and viral infections as well as with malignancies. The literature is very nonspecific regarding the types of malignancies connected with SJS and TEN, but this association may be related to the drugs used to treat specific malignancies or their side effects. Kasper (2001) observed an increased incidence of SJS in patients who were receiving both phenytoin and steroids and who completed whole brain irradiation for brain metastasis. Additional links to anticonvulsants and cranial irradiation are noted in the literature (Cockey, Amann, Reents, & Lynch, 1996; Micali, Linthicum, Han, & West, 1999).

SJS and TEN are immunologic in origin, but their exact etiology is not known (Patterson & Tripathi, 1999). Researchers have determined that apoptosis of keratinocytes occurs; this cell destruction is thought to be mediated or triggered by a cytokine, such as tumor necrosis factor. Antigen-antibody immune complexes are formed that trigger cytokine release. CD4 and CD8 T lymphocytes also are present in patients with SJS or TEN (Ellis et al., 2002; Kasper, 2001; Patterson & Tripathi).


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