Stevens-Johnson Syndrome and Toxic Epidermal Necrolysis: Oncologic Considerations

Keri Cassidy Hockett, ARNP, MSN, AOCN®

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.

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, sulfas, 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, & Daughtrey, 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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