Stevens-Johnson Syndrome

Stevens-Johnson syndrome (SJS) is a blistering skin disorder most often caused by drug reactions. It initially begins as a fever, but later involves the mucosa and skin, leading to blistering, necrolysis and skin sloughing. An important distinction to realize is that SJS involves less than 30% of the body surface area. If more than 30% of the body surface area is involved, it is classified as toxic epidermal necrolysis (TEN). This syndrome has a high mortality rate, but interventions include supportive care and intravenous immunoglobulin (IVIG) administration.

PLAY PICMONIC

Etiologies

Drugs

Med-bottles

The primary cause for SJS is adverse drug reactions. Some drugs associated with the development of SJS include antiepileptic medications, allopurinol, sulfa drugs, and penicillin.

Symptoms

< 10% of Body Surface Area (BSA) Involved

Less-than (10) Tin

SJS is distinguished from more severe blistering skin disorders by using the total amount of body surface area involved. If there is skin involvement in less than 10% of the BSA, then the disorder is classified as SJS.

10-30% BSA Overlaps with Toxic Epidermal Necrolysis (TEN)

(10) Tin to Dirty (30) with Toxic Skin Necrosis-crow

If 10-30% of the BSA is involved, there is an overlap of SJS and TEN. If greater than 30% of the BSA is involved, then the disorder is classified as TEN.

Fever

Fever-beaver

Initially, this disorder begins with fever. Patients begin to complain of fever, along with prolonged fatigue. Skin and mucosal involvement follows the initial presentation.

Affects Skin and Mucous Membranes

Skin-suit with Mucous

SJS affects the skin and mucous membranes leading to bullae formation and necrosis, along with sloughing of the skin. Typically two mucous membranes are involved (such as the eyes and mouth).

Bullae Formation

Bull-blisters

The skin pattern most commonly associated with SJS is widespread bullae formation. Bullae are large vesicles described as a rounded or irregularly shaped blisters containing serous or seropurulent fluid, equal to or greater than either 5 to 10 mm.

Necrosis

Necrosis-crow

SJS is characterized by confluent epidermal necrosis with minimal associated inflammation. Slow acetylators, patients who are immunocompromised, and those who are taking antiepileptic medications are at higher risk for necrosis because they can't completely detoxify reactive drug metabolites. These metabolites may present as antigens, leading to the production of tumor necrosis factor (TNF) alpha by the local tissue dendrocytes. This, in turn, causes epidermal cell apoptosis.

Sloughing of Skin

Skin Sloughing off

After epidermal necrosis, the skin begins to slough off in SJS revealing the dermal layer.

High Mortality Rate

High Death

This is a rare disorder, but has a high mortality rate of around 10%. However, if this progresses to toxic epidermal necrolysis (TEN), and involves more body surface area, the mortality rate increases to over 30%.

Treatment

Supportive Care

Supportive IV bags

Initial treatment for SJS is similar to that for patients with thermal burns, and care is only supportive. This involves intravenous fluids and analgesic administration, along with NG or parenteral feeding.

IVIG (Intravenous Immunoglobulin)

Ivy-gold-goblin

Intravenous immunoglobulin (IVIG) treatment is another promising treatment for reducing the length of the reaction and improving symptoms. This treatment modality has a higher success rate than corticosteroids.