Pathophysiology-Stevens-Johnson Syndrome/Toxic Epidermal Necrolysis

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Fever, malaise, fatigue and mucosal involvement, including ocular, cutaneous, severe, drug exfoliative syndrome and toxic epidermal necrolysis (SJS/TEN): could be a result of a genetic predisposition to drug hypersensitivity reactions marked by the destruction of keratinocytes and the epidermis, ending in necrosis and the widespread inflammation of the hypodermal layer, creating an acute, systemic Vitamin A toxicity. Granulysin, a cytotoxic protein produced in massive quantities by both CD8+ T lymphocytes and natural killer cells is the most prevalent molecule found in SJS/TEN biopsies. Granulysin is believed to act as a cytokine for destructive retinoid molecules (such as retinoid acid), combined together are believed to be responsible for keratinocyte apoptosis seen in SJS/TEN. As keratinocytes die off, the epidermis becomes detached from the dermis ending in tissue necrosis and sloughing (Teo & Walsh, 2016). Although not fully understood, the authors theorize that CD8+ T cells become hyperactive when exposed to extreme levels of retinol and overproduce Granulysin. Granulysin then attracts retinol acid which are both cytotoxic and together cause keratinocytes to die off. Keratinocytes make up 90% of the cells in the epidermis. Several combinations of Alleles and medications which when found that morbidity levels in patients often present with vague illness either through hepatic release as a result of liver injury, or through the inhibition of metabolism which leads to higher circulating retinol derivatives, such as retinoid acid, a powerful cell-lysing agent (Mawson, Erator, & Karre, 2015). Hepatic injury could explain the extended promodal illness seen in this disease as patients often present with vague illness which typically lasts for over a week prior to the onset of the more identifiable rash.

Implications: Pathophysiology
Understanding pathophysiology is key for treating and limiting the restorative mechanism for disease. Provided that retinal toxicity is involved with SJS/TEN, phasmapheresis could be used to reduce circulating levels and mitigate the destructive effects which retinol contributes in this disease. Plasmapheresis has been shown in small samples to reduce mortality versus typical treatment for patients with SJS/TEN (Mawson, 2015).

Genetic predispositions have been positively established for people of Asian descent who carry the HLA-B1502 Allele when exposed to Carbamazepine. The FDA recommends genetic testing prior to starting Asian patients on Carbamazepine therapy (Tangamansorn, Chalyakunapruk, Sorinka, Lohitnavy, & Tassaneeyakul, 2013). Studies have also linked more disease severity for patients with SJS/TEN who are positive for the B*1502 Allele and Carbamazepine (Patel, Eriator, & Karre, 2015) especially during debridement and dressing changes will be needed. Patients at risk should familiarize themselves with standardization for immune system reconstitution. Nursing: Implications:

Rapid identification of disease is crucial for patients suffering from SJS/TEN as removal of the offending medication is critical for mitigating the effects of the illness. Nursing should familiarize themselves with the symptoms and signs of progression as illness may advance quickly from SJS to TEN in as few as twelve hours (Poulson, Nielsen, & Poulsen, 2013). Patients should be warned of signs or symptoms of decompensation to respiratory failure or sepsis. Pain management and the use of conscious sedation, especially during debridement and dressing changes will improve healing and health outcomes. Treatment is best done in a specialized burn center. The nurse should seek to coordinate a multi-disciplinary team approach (Cooper, 2012). Long-term restrictions for this disease is frequently seen with SJS/TEN and patients will often require long-term use of bronchodilators (British Journal of Medicine, 2015).

References

Stevens-Johnson Syndrome and Toxic Epidermal Necrolysis
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Introduction
Stevens-Johnson Syndrome (SJS) and Toxic Epidermal Necrolysis (TEN) are rare, cutaneous, severe, drug-induced hypersensitivity reactions marked by widespread inflammation of the epidermis, ending in necrosis and the eventual sloughing of tissue. First described in 1922 by pediatricians Albert Stevens and Frank Johnson, both diseases are believed to be a result of the same disease process. They are the most serious of all drug-related hypersensitivity reactions which result in hospitalization (Ferrandiz-Perez & Garcia-Patos, 2013). As a critical care nurse caring for a variety of trauma patients, it is crucial that staff is aware of potential serious medication reactions. Some of the most serious of these start out with vague symptoms and may appear benign. However, some are very serious and potentially fatal.

Signs and Symptoms
Early signs/symptoms:
• Fever, malaise, fatigue and mucosal lesions, headache, bleeding
Later signs/symptoms:
• Marked erythema of skin leading to papules, vesicles and redness. These start on the face, neck and anterior trunk and may extend over the entire surface of the skin.
• Mucosal involvement, including ocular, GI, GU, genital and upper and the epithelial cells of the lower respiratory tract. These areas are associated with heavy bleeding, scarring and long-term complications.

Statistics
Disease TBSA % Mortality Incidence/million
SJS <10% 5% 2-6
TEN >30% (Mawson et al, 2015) >30% 30% 0.2-1.2
(Mawson et al, 2015) >10%, >80% is considered to be read SJS/TEN disease) (TBSA-total body surface area)