

STEVENS-JOHNSON SYNDROME (L55.1)

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REMEMBER

- ▶ Steven-Johnson Syndrome (SJS) is a potentially life-threatening drug reaction with mucocutaneous erythema, and dermo-epidermal detachment.
- ▶ The disease generally occurs in adults, but it can also affect the pediatric population. The possibility of Hodgkin's disease should not be forgotten in the pediatric SJS/toxic epidermolysis (TEN).
- ▶ SJS and TEN are a spectrum of a disease. It is evaluated according to the involvement area in the skin. It is defined as SJS <10%, SJS-TEN overlap between 10-30%, TEN >30% epidermal detachment.
- ▶ Steven-Johnson Syndrome should not be considered as a skin-limited disease, but a serious systemic disease. The patients should be followed up in the burn unit, if possible.
- ▶ Steven-Johnson Syndrome/toxic epidermolysis can cause serious complications in the pulmonary, renal and hepatic systems, which can be fatal.
- ▶ Most common culprit drugs associated with etiology: allopurinol, non-steroidal anti-inflammatory drugs (NSAIDs), antibiotics (especially sulfonamides), and antiepileptics (e.g., carbamazepine).
- ▶ The disease may also be triggered by Herpes simplex virus and Mycoplasma infections.

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Nephrology

- ▶ Patients with SJS may develop acute renal failure (ARF) due to intense trans-epidermal fluid loss. In the presence of increased serum urea and creatinine, patients should be consulted with nephrology for the treatment of ARF.
- ▶ Some cases have been reported to progress to chronic renal failure.

Infectious Diseases

- ▶ Patients are at high risk of infection due to impaired skin integrity. If infection is added to the existing picture, it may progress to sepsis.
- ▶ Sepsis/septic shock is the leading cause of death in these patients. In order to prevent progression to sepsis, an infectious diseases specialist should be consulted for the need for appropriate systemic antibiotic therapy in the early stage.

Nutrition and Dietetics

- ▶ There is intense protein loss in patients due to deterioration of skin integrity, and increased catabolism. There may also be feeding difficulties due to oral involvement. These patients should be consulted with a dietitian for appropriate nutritional support.

Psychiatry

- ▶ Individuals with the disease have an increased risk of major depression and post-traumatic stress disorder. Psychiatric evaluation before the discharge of the patients is strongly recommended.

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STEVENS-JOHNSON SENDROMU (L51.1)

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HATIRLA

- Steven-Johnson sendromu (SJS) mukokutanöz eritem ve dermo-epidermal ayrışmanın izlendiği, ölümcül olabilen bir ilaç reaksiyonudur.
- Genellikle erişkinlerde görülmekle beraber pediatrik yaş grubunda da ortaya çıkabilir.
- SJS ve toksik epidermoliz (TEN) bir spektrumdur. Derideki ayrışmaya göre değerlendirilir. SJS <%10, SJS- TEN overlap %10-30 arası, TEN>%30 epidermal ayrışma olarak tanımlanır.
- Steven-Johnson sendromu sadece bir deri hastalığı olarak görülmemeli ve ciddi bir sistemik hastalık olarak, mümkünse yanık ünitesinde takip edilmelidir.
- Steven-Johnson sendromu/ toksik epidermoliz pulmoner, renal ve hepatik sistemde ölümcül olabilecek ciddi komplikasyonlara neden olabilir.
- Etiyolojide en çok suçlanan ilaçlar: allopürinol, steroid olmayan anti-inflamatuvar ilaçlar (NSAİİ'ler), antibiyotikler (özellikle sulfanomid grubu) ve antiepileptiklerdir (ör: karbamazepin).
- Herpes simpleks virüs ve Mycoplasma enfeksiyonları tarafından da SJS tetiklenebilir.
- Radyoterapi sonrasında da SJS/TEN tablosu ortaya çıkabilir.
- Bazı HLA tipleri (HLA-B*15:02) SJS/TEN hastalıkları için yatkınlık oluşturmaktadır.

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tabloya enfeksiyonun eklenmesi halinde sepsise ilerleyebilir.

- ▶ Sepsis/septik şok bu hastalarda başlıca ölüm nedenidir. Sepsise ilerlemenin önlenmesi için erken dönemde uygun sistemik antibiyoterapi ihtiyacı açısından enfeksiyon hastalıklarına danışılmalıdır.

Beslenme ve Diyetetik

- ▶ Deri bütünlüğünün bozulması ve artan katabolizma nedeniyle hastalarda yoğun protein kaybı mevcuttur. Aynı zamanda oral tutuluma bağlı beslenme güçlüğü de olabilir. Bu hastalar uygun nutrisyonel destek amacıyla diyetisyene danışılmalıdır.

Psikiyatri

- ▶ Hastalığı geçiren bireylerde majör depresyon ve post-travmatik stres bozukluğu riski artmıştır. Taburculuk öncesi psikiyatrik görüşme mutlaka önerilir.

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