MODERN APPROACHES TO THE MANAGEMENT OF PATIENTS WITH STEVENS-JOHNSON SYNDROME AND TOXIC EPIDERMAL NECROLYSIS.

T. R. Umanets[1], A. A. Buratynska[1], V. E. Khomenko[2], S. V. Zaikov[3]

[1]State Institution «Ukrainian Center of Maternity and Childhood of the National Academy of Medical Sciences of Ukraine», Kyiv, Ukraine
[2]Bogomolets Medical University, Kyiv, Ukraine
[3]Shupyk National Healthcare University of Ukraine, Kyiv, Ukraine

Abstract. The article presents updated information on Stevens-Johnson syndrome (SJS) and toxic epidermal necrolysis (TEN) also known as Lyell's syndrome, which are severe cutaneous non-IgE-mediated hypersensitivity reactions characterized by intense necrosis and detachment of the epidermis with the formation of large blisters and erosions on the skin and mucous membranes. The cause of SJS and TEN can be such medications as anticonvulsants and antiepileptics (phenytoin, carbamazepine, phenobarbital, valproic acid and its derivatives, lamotrigine); antibiotics (ampicillin or amoxicillin, fluoroquinolones, cephalosporins, penicillin); antimicrobials (trimethoprim, sulfonamides, sulfamethoxazole); nonsteroidal anti-inflammatory drugs (piroxicam, meloxicam, ibuprofen, acetaminophen); other drugs (allopurinol, chlormezanone), etc. In the pathogenesis of these conditions, the leading role is played by the immune complex reaction of type III hypersensitivity or the immune reaction of delayed hypersensitivity of type IVc, which leads to cytotoxic apoptosis of keratinocytes mediated by CD8+ T-cells. The diagnosis of SJS and TEN is based on the history and clinical features. At least 3 of the following criteria are required for diagnosis: 1) presence of purpuric macules or atypical targets that are not predominantly distributed on the extremities; presence of vesicles, bullae; 2) epidermal detachment (appearance of "wet" clothing); 3) positive Nikolsky sign; 4) multifocal mucosal erosions (enanthema, bullae, erosions in the oral cavity, nasopharynx, oropharynx, nose, eyes, or genital/anal area). Laboratory tests may be useful in assessing multisystem involvement and the severity of the patient's condition. The main goal of treatment is to reduce the development and progression of complications. It is necessary to immediately exclude the suspected drug, which is identified using the ALDEN algorithm. First of all, treatment should be aimed at monitoring vital signs, maintaining thermoregulation and pain control (analgesia), consuming sufficient fluids, preventing fluid loss and electrolyte imbalance, and preventing secondary infection, the development of long-term complications, and scarring.

In conclusion, SJS and TEN (Lyell's syndrome) are severe multisystem hypersensitivity reactions triggered by specific drugs. Diagnosis and treatment of these conditions are complex and require an interdisciplinary approach. Increasing the availability of pharmacogenetic screening (HLA) opens up possibilities for the prevention of SJS/TEN in individuals prone to their development. The prospects for the use of immunomodulatory agents in SJS/TEN to increase the effectiveness of treatment, prevent the development of complications, and reduce mortality also require further study.

Key words: Stevens-Johnson syndrome, toxic epidermal necrolysis, epidemiology, etiology, pathogenesis, clinical manifestations, diagnostics, treatment.