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НАУЧНО-ПРАКТИЧЕСКИЙ ЖУРНАЛ

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DIFFERENTIAL DIAGNOSIS OF TOXIC-ALLERGIC BULLOSIC EPIDERMAL NECROLIS AND STEVENS-JOHNSON SYNDROME IN CHILDREN

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Toxic epidermal necrolysis or Lyell's syndrome (SL, TEN) is the most severe form of drug-induced toxicoderma [1]. Named in honor of the Scottish dermatologist Alan Lyell, who first described this disease in 1956.

Toxic epidermal necrolysis (TEN) develops as a reaction to the combined effects of toxic, medical and infectious agents, which appears as a high degree of body hypersensitivity [2].

The most common causes of toxic epidermal necrolysis are: sulfa drugs, antibiotics (erythromycin, penicillins, tetracyclines, streptomycin, etc.), barbiturates, analgesics (phenylbutazone, salicylates, paracetamol, piroxicam, diclofenac, cephalosporins, fluoroquinolones, etc [6].

Other causes of Lyell's syndrome may be chemical intoxication or tainted food.

Stevens-Johnson syndrome (SSD) is an acute bullous lesion of the mucous membranes and skin of an allergic reaction, characterized by extensive lesions of the skin and mucous membranes, induced by medication [2,15]

Data on Stevens-Johnson syndrome were published in 1922. Over time, the syndrome was named after the authors who first described it. The disease is a severe form of multiforme exudative erythema known as - "malignant exudative erythema".

Together with Lyell's syndrome, pemphigus, bullous variant of SLE, allergic contact dermatitis, Hailey-Hailey disease, etc. Stevens-Johnson syndrome is referred as bullous dermatitis, the common clinical symptom of which is the formation of blisters on the skin and mucous membranes.

Stevens-Johnson syndrome occurs at any age, most often in persons 20-40 years old and extremely rare in the first 3 years of a child's life. According to various data, the prevalence of the syndrome per 1 million population ranges from 0.4 to 6 cases per year.

Diagnosis of Stevens-Johnson syndrome is based on the severe course of the disease and the development of rashes which are characteristic for bullous multiform exudative erythema, not only on the skin, but also on mucous layer of the oral cavity, around the external opening of the urethra, in the anal-genital region, on the conjunctiva.

Depending on the area of the affected skin, the following forms of ELR are distinguished: Stevens-Johnson syndrome (SSD) - less than 10% of the body surface; toxic epidermal necrolysis (TEN, Lyell's syndrome) - more than 30% of the body surface; intermediate form of SJS / TEN (10-30% of the skin is affected).

Lyell's syndrome is characterized by a large affected surface of the skin. A feature of this disease is the exfoliation of the epidermis (a symptom of Nikolsky) [4].

Nikolsky P.V. was the first to describe the symptom of epidermal detachment in pemphigus in 1886. Nikolsky's symptom is a clinical manifestation of acantholysis, which in pemphigus vulgaris can be positive both in the lesion and near it, as well as on apparently healthy skin away from the lesion. With a symptom of Nikolsky, the skin peels off as if burned with boiling water, with pemphigus vulgaris it will be positive in the lesion and rarely on healthy skin (when rubbing on skin that seems healthy, there is a slight rejection of the upper layers of the epidermis). After exfoliation of the epidermis, erosions form in its place, which bleed and are sharply painful on palpation. Rashes and erosion can be on the mucous membrane of the oral cavity, pharynx, esophagus, genital organs. Stevens-Johnson syndrome is often difficult to distinguish from the initial form of SL [6,12,17].

The decisive differential diagnostic criteria are the development of epidermal necrolysis and a positive Nikolsky symptom in Lyell's syndrome, which is not typical for SJS.

In modern terminology, both syndromes are combined into a common nosology - Stevens-Johnson syndrome - toxic epidermal necrolysis (SSD-TEN) [2,3].

We conducted an expert evaluation of 3 case histories in sick children treated in the ICU department of the 1st clinic of the Tashkent Medical Academy for severe allergic lesions of the skin and mucous membranes in the form of Lyell and Stevens-Johnson syndrome for the period 2022-2023. Two children (aged 7 months, 2 years and 5 years old) had SL, one had SJS (child 11 years old). It was found that two children had a history of the appearance of rashes after the use of medications (ceftriaxone, gramox) [4]. The rest of the children had errors in nutrition (they used citrus fruits). All four children were admitted to the department in a serious condition, with fever, intoxication, and various elements of skin allergic manifestations and rashes on the mucous membranes in terms of morphology and prevalence. In principle, the scheme of treatment of children did not differ: antibiotics - cephalosporins of the 3rd generation and amikacin (in a child with an allergic reaction to ceftriaxone) were used to prevent infectious complications. All children underwent hormone therapy with glucocorticoids at a daily dose of dexamethasone 1-2 mg/kg/day. [3,5,14].

Complications of Toxo epidermal necrosis syndrome are a progressive deterioration in the general condition of the patient and becomes extremely severe in a short period of time. Excruciating thirst, decreased sweating and saliva production are signs of dehydration.

Thus, we can conclude that none of the children from SJS had a positive Nikolsky symptom. At the same time, in children diagnosed with Lyell's syndrome, the severity of the lesion with generalization of bullous epidermolysis, the extent and tendency to merge erosions were noted [8, 17]. There is severe pain in the area of the entire skin and mucous membranes. The prognosis largely depends on the degree of damage, the presence of infectious complications, the timeliness and volume of medical care provided. The average mortality rate is 25-30%, in severe cases it can reach 65-70%. All this emphasizes the commonality of the etiology, pathogenesis, and management of patients with SL and SJS. [11].

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