CHURG-STRAUS SYNDROME IN CHILDREN: CASE REPORT

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Churg-Straus syndrome (CSS) is rare disease in children. At the moment, only several decades of observations are described. However, due to severe course and unfavorable prognosis, it attracts focused attention of specialists. CSS is systemic disorder which is characterized with severe bronchial asthma, pulmonary infiltrates, hypereosinophilia and systemic vasculitis. Frequency of CSS in adults is 2.4 /1 mln/year; data regarding its frequency in children are absent due to its rarity. This disease manifests with allergic rhinitis, bronchial asthma, eosinophilic infiltrative disease, and systemic vasculitis with granulomatous infiltration. Outcome is fatal if not treated. Predominant causes of death are cardiac, renal, cerebral failure, and perforation or bleeding in gastrointestinal tract. Diagnostic criteria of SCS proposed by the American College of Rheumatology are: bronchial asthma, eosinophilia, mono- or polyarthropathy, affection of accessory sinuses, eosinophilic extravasates on biopsy.

A clinical case of CSS is presented. A girl, 11 years old, suffered from quickly progressive bronchial asthma on the background of hypereosinophilia and elevated IgE level.

Quickly progressive bronchial obstruction syndrome with unusual course appeared:

- with abundant passage of mucous-purulent sputum
- with local fine crackles and crepitation predominantly in basal and lower parts which appeared and disappeared during the day
- with catarrhal-purulent diffuse endobronchitis on bronchoscopy

A large quantity of eosinophils was found in sputum (up to 80%). In sputum, bacterial culture fungi of Candida spp. and streptococci in diagnostic significant titers were revealed, IgE 710 mcmol/l. Preliminary diagnosis: bronchial asthma. Infiltrative changes in lungs on X-ray and CT scan were detected. In biopsy of skin and muscles from the back and left foot, systemic productive vasculitis, granulomatous inflammatory infiltration and muscular calcification were revealed allowing to diagnose Churg-Strauss systemic vasculitis.

Lethal outcome occurred due to perforative enteritis, perforation of esophagus, generalized peritonitis and cardiac-pulmonary failure.

Summary. The rarity of the pathology (1–3 cases/1 000 000)makes it difficult to diagnose. Nevertheless, in presence of bronchial asthma, eosinophilia and affection of inner organs, it is necessary to keep in mind Churg-Strauss vasculitis. Latest developments of immunomodulatory therapy and early

beginning of the treatment allow to improve outcome of the disease