OVERWHELMING POSTSPLENECTOMY INFECTION SYNDROME IN CHILD WITH AUTOIMMUNE LYMPHOPROLIFERATIVE SYNDROME: A CASE REPORT

O. Samodova, E. Krieger

Northern State Medical University, Arkhangelsk, Russia

Overwhelming postsplenectomy infection (OPSI) syndrome is a life-threatening condition that occurs in splenectomized patients. Asplenic patients are at increased risk of generalized infections due to disorders of opsonization and phagocytosis of encapsulated bacteria.

A six year-old girl, who had splenectomy for hypersplenism at the age of two years, was admitted to an intensive care unit with septicemia and septic shock. The patient had a history of primary immunodeficiency, autoimmune lymphoproliferative syndrome. She had been vaccinated with the polyvalent pneumococcal vaccine in two years after splenectomy.

The first symptoms of the disease were nonspecific and included febrile fever, headache and nausea. Haemorrhagic rash arose on the third day of the illness. Physical examination revealed a toxemic appearance, presenting with fever, mottled skin, acrocyanosis, slow recoloration time, tachycardia and hypotension. Meningeal irritation findings were positive. Petechiae were localized on the face, trunk, extremities and oral mucosa. Laboratory tests revealed trombocytopenia with disseminated intravascular coagulation. Streptococcus pneumoniae were isolated from blood and normal cerebrospinal fluid.

Treatment comprised antibiotics, intravenous fluids, steroids, heparin, immunoglobulins. The patient was discharged from hospital with recovery.

The case demonstrated the successful experience of OPSI treatment in immunocompromised asplenic patient with primary immunodeficiency. We would like to emphasize that in case of necessary splenectomy, the patient should be given pneumococcal vaccine before surgery to reduce the risk of subsequent OPSI.