Pediatric Rheumatology

Oral presentation

14.2 Causes of early death in juvenile onset systemic lupus erythematosus (JSLE)

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Purpose

To describe the causes of death occurring within the first month following the diagnosis of JSLE in order to prevent them.

Methods

Retrospective study of causes of early death in JSLE patients during the period 1995 – 2006 conducted on behalf of Paediatric Rheumatology International Trials Organization (PRINTO). All the patients fulfilled the American College of Rheumatology (ACR) criteria for the diagnosis of SLE, and were diagnosed before 16 years of age.

Results

Death was recorded in 5 girls and 1 boy including three non Caucasian patients, aged 6 to 16 years. Initial SLE manifestations comprised at least three organ involvement, including at least two major organ involvement: central nervous system (4 patients), kidney (5 patients), pancreas (2 patients), hematopoietic system (3 patients), heart (1 patient), skin (2 patients), and joints (3 patients). Despite administration of oral and pulsed steroids, associated twice with intravenous cyclophosphamide, SLE activity remained uncontrolled in all the patients. Death resulted from SLE organ failure either alone in 2 patients (pancreatitis: 1 patient, neurolupus: 1 patient), either associated with thrombotic event (catastrophic antiphospholipid syndrome: 2 patients, pulmonary thromboembolism: 1 patient with nephrotic syndrome) and/or infection (paravertebral abcess: 1 patient, pneumococcal sepsis: 1 patient).

Conclusion

Despite prompt diagnosis and management, death may occur at presentation of juvenile-onset SLE. Half of them resulted from thromboembolic event in patients with nephrotic syndrome or APL. These features suggest that prompt prophylactic anticoagulation may be beneficial in patients with severe SLE multiorgan involvements associated with risk factors of thromboembolic event.