Ecthyma gangrenosum as the presenting clinical feature of X-linked agammaglobulinaemia: report of three cases and a review of literature

ABSTRACT

Background: Children with X-linked agammaglobulinaemia (XLA) usually present with pneumonia and otitis media caused by pyogenic bacteria. Rarely, ecthyma gangrenosum (EG), a known cutaneous manifestation of Pseudomonas septicaemia present in XLA as the first presenting features. We report three cases of EG caused by P. aeruginosa in previously healthy boys, leading to the diagnosis of XLA. In addition, we provide a brief literature review on those cases of EG where an underlying XLA was eventually diagnosed. Methods: Three paediatric cases admitted to the intensive care unit with P. aeruginosa septicaemia associated with ecthyma gangrenosum were reviewed retrospectively. Laboratory workup consisted of microbiological, haematological and immunological investigations were analysed. Results: The ages of the three patients were: one year and six months, three years and five months, and five years and six months. All patients had septic shock and required mechanical ventilation. P. aeruginosa was isolated in the blood and/or skin lesions of all patients. Underlying hypogammaglobulinaemia and neutropaenia were detected in all patients. Treatment consisted of combined antipseudomonal antimicrobial therapy and surgical debridement. All patients survived. Subsequent B-cell measurement and Bruton's tyrosine kinase (BTK) protein and genetic analysis confirmed the diagnosis of XLA. Twelve other similar reported cases were reviewed and analysed based on their clinical presentation, diagnosis and treatment. Conclusion: P. aeruginosa sepsis should be treated as early as possible. The most common risk factor for ecthyma gangrenosum in XLA patients is neutropaenia. In previously healthy children presenting with EG, immunological evaluation is important to rule out an underlying immunodeficiency.

Keyword: Ecthyma gangrenosum; Pseudomonas aeruginosa; X-linked agammaglobulinaemia; XLA; Neutropaenia