

Ecthyma gangrenosum as the presenting clinical feature of x-linked agammaglobulinemia: Report of three cases

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ABSTRACT

Background: Children with x-linked agammaglobulinemia (XLA) usually presents with pneumonia and otitis media caused by pyogenic bacteria. Rarely, ecthyma gangrenosum (EG), a known cutaneous manifestation of *Pseudomonas* septicemia present in XLA as the first presenting features. We report three cases of EG caused by *Pseudomonas aeruginosa* in previously healthy boys, leading to the diagnosis of XLA. Additionally, 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* septicemia associated with ecthyma gangrenosum were retrospectively reviewed. Laboratory workup consisted of microbiological, hematological and immunological investigations. **Results:** The three patients were aged 1 year 6 months, 3 years 5 months and 5 years 6 months. All patients had septic shock and required mechanical ventilation. *Pseudomonas aeruginosa* was isolated in blood and/or skin lesions of all patients. Underlying hypogammaglobulinemia and neutropenia were detected in all patients. Treatment consisted of combined antipseudomonal antimicrobial therapy and surgical debridement. All patients survived. Subsequent B cells measurement and 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 neutropenia. In previously healthy children, immunological evaluation is important to rule out an underlying immunodeficiency.