Campylobacter jejuni Infection in X-linked Agammaglobulinemia Patients

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ABSTRACT

XLA (X-linked agammaglobulinemia patients) is a relatively rare form of immunodeficiency characterized by a profound deficiency of B-lymphocytes and recurrent infections. Most clinical problems are recurrent sinusitis, pneumonia, otitis media from encapsulated bacteria such as Haemophilus influenza and Streptococcus pneumoniae. There are case reports of Campylobacter jejuni bacteremia which is rare in immunocompetent patients. Other rare complications in patients with XLA and campylobacter bacteremia such as campylobacter pericarditis, myocarditis, myopericarditis, and chronic skin infection has been reported. Treatment of Campylobacter in patients with XLA in case reports have varied in durations. Most require a prolonged period of antibiotic treatment with a mean duration of approximately 10 months and adequate immunoglobulin levels from intravenous immunoglobulins (IVIG). Despite immunoglobulin substitution therapy with normal IgG levels, severe infections with Campylobacter and Helicobacter can relapse and emerge. This is suspected from inadequate IgA and IgM in IVIG substitution and the capability of intracellular harbor in gastrointestinal tract. Interruption of transmission from poultry by avoidance of unpasteurized dairy is a major intervention in preventing human Campylobacter infection.

In conclusion, C. jejuni requires humoral immunity to confine infection and patients with XLA are prone to these infections. These organisms are not clearly stained by Gram stain and are also fastidious, so culture is also difficult. The diagnosis should be kept in mind when treating the patient with XLA. (J Infect Dis Antimicrob Agents 2015;32:69-75.)

INTRODUCTION

XLA (X-linked agammaglobulinemia patients) is a relatively rare form of immunodeficiency characterized by a profound deficiency of B-lymphocytes. This deficiency is attributable to an arrest in B-lymphocyte development caused by a defect in a tyrosine kinase, named Bruton tyrosine kinase. The gene that is responsible for this Bruton tyrosine kinase is mapped to X chromosome, in the Xq22 region. It is an X-linked recessive disease but about one-third of XLA cases are sporadic. Patients with XLA have few immunoglobulins because plasma cells are depleted. Despite this defect, most patients still have some immunoglobulins in their serum because there are a small percentage of B-lymphocytes that are able to escape the block in

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