X-linked agammaglobulinaemia (XLA), also known as Bruton’s agammaglobulinaemia, is characterized by low or negligible levels of serum immunoglobulins (1). XLA is caused by mutations in the gene coding for Bruton’s tyrosine kinase (BTK). Deficiency of BTK leads to a developmental block in B-cell differentiation; hence, patients essentially lack antibody-producing plasma cells and are susceptible to various infectious diseases (2). Infections with unusual organisms, such as Helicobacter cinaedi, may also be troublesome (3). H. cinaedi, previously known as Campylobacter cinaedi, has been frequently reported in HIV-positive patients (4). In immunocompromised patients, the spectrum of illnesses caused by H. cinaedi includes recurrent fever, bacteraemia, arthritis, osteomyelitis, cellulitis, abdominal abscesses and diarrhoea. However, signs of inflammation, such as fever, may be delayed in immunocompromised patients, possibly due to the absence of functional antibodies required for opsonization and phagocytosis (3). A few cases of XLA with chronic infections, leg cellulitis and pyoderma gangrenosum-like ulcer, caused by H. cinaedi have been reported, although common Helicobacter species have been recognized as pathogens in many more patients with XLA (3, 5, 6). In this report, we describe a patient with XLA complicated by recurrent cellulitis caused by H. cinaedi.

CASE REPORT

The proband is a 38-year-old Japanese man with agammaglobulinaemia that was diagnosed in infancy. He had been treated with pH4-treated acidic normal human immunoglobulin. He had had recurrent episodes of feverish, painful swelling and erythemas on the dorsum of the right hand, the left lower leg and the left ankle since the age of 36 years. On one occasion, the feverish swelling and erythemas accompanied by pain on the left lower leg was also accompanied by high fever and elevation of circulating C-reactive protein at 2.36 mg/dl (normal range: 0–0.3 mg/dl) and he was hospitalized. The results of laboratory tests were as follows: white blood cell (WBC) count 8,400/mm$^3$ (normal 3,800–8,500/mm$^3$); IgG 1,169 mg/dl (870–1,700 mg/dl) under treatment with pH4-treated acidic normal human immunoglobulin; IgA < 1 mg/dl (110–410 mg/dl); and IgM < 1 mg/dl (35–220 mg/dl). One week after the onset of swelling, the erythemas developed into brown macules (Fig. 1A, B). A skin biopsy from the left lower leg revealed slight fibrosis and mild lymphocytic infiltration in the upper dermis. Repeated cultures for microorganisms from venous blood samples detected a spirillum (Fig. 1C). No bacterial culture or PCR tests were performed on the skin samples. H. cinaedi was confirmed by PCR using the specific primers for the 16S rRNA of H. cinaedi (7) and by mass spectrometry (MS) with Vitek MS (BioMérieux, Durham, NC, USA), using a matrix-assisted laser
H. cinaedi in chronic skin infections cause by unusual bacteria. Our results suggest the importance of checking bacteraemia carefully by the appropriate techniques in immunocompromised patients, such as those with XLA.

The authors declare no conflicts of interest.

REFERENCES