

Bilateral lower leg swelling and lymphocytic panniculitis in a patient with XLA

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PA is a 23 year old male who was diagnosed with X-linked agammaglobulinemia (XLA) at the age of 7 year after a long history of recurrent fevers and persistent sinusitis. Since then he has been on IVIG replacement and has had only a few infections, which included two episodes of pneumonia treated as an outpatient and 2 episodes of sinusitis.

At the end of 2002, he presented with tender, warm, blanching nodules, on the anterior aspect of his left tibia. Subsequently, those nodules coalesced in a brownish, indurated lesion with fibrotic edges. Two skin biopsies, done one year apart, were consistent with erythema nodosum. He was briefly treated with potassium iodide, with only minor improvement.

In June, 2003 he started to develop scattered erythematous, indurated 2 cm skin lesions, on his arms and flanks and legs, that typically lasted a few days and resolved spontaneously without scarring. A biopsy was consistent with panniculitis. In June 2004 he developed bilateral leg swelling, pain and intermittent low grade fever. In August 2004 he began to have occasional ulceration of the edematous/fibrotic lesion in the anterior aspect of his left lower leg, which was very painful and typically healed after 2-3 weeks. On some occasions, the ulcers resolved spontaneously; other ulcers cleared after treatment with Zithromax and Naprosyn. Attempts to treat with Dapsone and minocycline were unsuccessful. He developed bilateral inguinal lymphadenopathy. A bone marrow biopsy and lymph node biopsy did not reveal a malignancy. CBC's have shown high WBC (15-22,000 range), with marked neutrophilia (12-18,000) and high ESR (50-70).

He was lost to follow up for a period of 3 months, then presented acutely with several purulent ulcers and swelling of his lower extremities. CT scan and MRI of the legs showed bilateral cellulitis involving the deep tissue. Numerous blood cultures were negative. A wound culture grew enterococcus sensitive to beta lactams and vancomycin. After 7 days of vancomycin treatment, the legs ulcers didn't show any improvement, and he developed persistent fever (39-40 C), despite negative blood culture. An empiric treatment with Zosyn was started. He defervesced in 18 hours. 48 hours after Zosyn the round scattered lesion of panniculitis disappeared. After one week of zosyn, all the ulcers improved significantly. All the ulcers healed after 3 weeks of treatment with Zosyn.

Labs at admission revealed (IgG 880 mg/dL, IgA<5, IgM<5, and IgE<4). CBC showed WBC of 22,000 (80% neutrophils, 20% lymphocytes), Hct 26%, Hgb 9.8, ESR 70, CRP 22. After 1 week of Zosyn CRP was 1.41, ESR 2, WBC 6,250 (60% neutrophils and 28% lymphocytes).

Skin infection and cellulitis from gram negative organisms, often localized on the lower extremities, have been described in XLA patients. Gram negative agents, sometimes

atypical and difficult to culture, such as *Campylobacter jejuni* and *Flexispira* spp have been reported in the literature. Empiric IV antibiotic treatment might be necessary in such patients even if blood cultures and skin cultures are negative, or non-diagnostic.