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Unmasking Immune Reconstitution Inflammatory Syndrome: Nontuberculous Mycobacterial Scrofula

To the Editor:

A 29-year-old man with a history of human immunodeficiency virus (HIV) presented with a left neck mass. He was diagnosed with HIV 6 years previously but went untreated until 6 weeks earlier when he developed thrush and was found to have a CD4 count of 13 cells/mm^3 and an HIV-1 viral load of 1.15 $\times$ 10^6 RNA copies/mL. At that time, antiretroviral therapy consisting of tenofovir-emtricitabine and ritonavir-boosted atazanavir was initiated. After 4 weeks on antiretroviral therapy, his CD4 count and HIV viral load improved to 182 cells/mm^3 and 1640 RNA copies/mL, respectively; however, he developed an enlarging nontender left-sided neck mass (Figure 1A). Computed tomography revealed extensive left greater than right centrally necrotic cervical lymphadenopathy, with the largest node in the left cervical chain measuring 8 cm in diameter (Figure 1B). Fine-needle aspiration revealed 3 to 4+ acid-fast bacilli with the state laboratory subsequently confirming *Mycobacterium avium-intracellulare* complex by polymerase chain reaction testing. He was treated for *M. avium-intracellulare* with triple therapy consisting of clarithromycin, ethambutol, and rifabutin. He was prescribed a 4-week prednisone taper for presumed immune reconstitution inflammatory syndrome in addition to continuing with antiretroviral therapy. After hospital discharge, the mass spontaneously drained cutaneously, and within weeks his neck swelling had resolved.

Immune reconstitution inflammatory syndrome is an exaggerated inflammatory response that occurs within 3 months of antiretroviral therapy initiation in patients with acquired immunodeficiency syndrome corresponding to immune recovery as evidenced by a sharp increase in the CD4 T-cell count and decline in the HIV viral load. Immune reconstitution inflammatory syndrome occurs in approximately 11% of patients initiating effective antiretroviral therapy and is more common in patients with initial CD4

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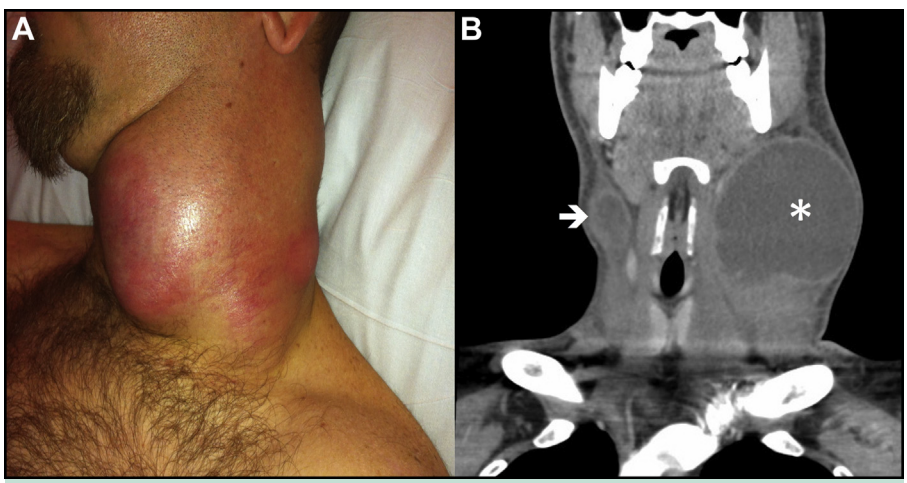

Figure 1 (**A**) Photograph of patient on presentation with large nontender left-sided neck mass. (**B**) Neck computed tomography scan showing left (*asterisk*) greater than right (*arrow*) centrally hypoattenuating cervical lymphadenopathy. The left cervical lymph node shown measures approximately 8 cm in diameter.

counts of less than 50 cells/mm^3.[1,2] Immune reconstitution inflammatory syndrome is often deemed paradoxical or unmasking. Paradoxical immune reconstitution inflammatory syndrome refers to a paradoxical worsening of a known opportunistic infection despite appropriate treatment. Unmasking immune reconstitution inflammatory syndrome refers to a new presentation of an opportunistic infection that is "unmasked" after starting antiretroviral therapy.[2] The incidence of nontuberculous mycobacterial immune reconstitution inflammatory syndrome is approximately 3.5% in patients initiating antiretroviral therapy with CD4 counts less than 100 cells/mm^3. Of these patients, nontuberculous *M. avium-intracellulare* peripheral lymphadenitis is the most common clinical presentation, seen in approximately one third of cases, with approximately one third of these primarily involving the cervical lymph nodes.[3] Mycobacterial cervical lymphadenitis (also known as "scrofula") is associated with both tuberculosis and nontuberculous mycobacterial infections and presents as a chronic, painless neck mass that grows with time and can rupture and drain spontaneously. The majority (~90%) of nontuberculous mycobacterial immune reconstitution inflammatory syndrome cases have a clinical response to systemic glucocorticoid therapy.[3]

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