

Headache and Rash in a Healthy Adult Male

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2. Clinical Image

A 50-year-old male with a past medical history of psoriasis presented with a two-week history of progressively worsening fatigue, low-grade fevers and throbbing frontal headaches, and several ulcerated papules of varying size (1-5mm) with central crusting and surrounding erythema noted on the forearms, chest, back and feet (**Figure 1**). A lumbar puncture (LP) demonstrated an elevated opening pressure and detection of cryptococcal antigen in the cerebral spinal fluid (CSF). HIV-screen was repeatedly negative. Magnetic resonance imaging (MRI) of the brain revealed cerebral atrophy.

This case should raise suspicion for disseminated cryptococcosis. Its rash frequently presents as molluscum contagiosum-like umbilicated papules of varying sizes, but many other manifestations (including vegetating crusted plaques or ulcers) have also been visualized. Disseminated cryptococcosis is rare in the absence of acquired immunodeficiency syndrome (AIDS) or use of immunosuppressive agents, and usually suggests an underlying primary or secondary immunodeficiency syndrome. Quantification of CD4⁺ lymphocyte count can reveal a rare but severe disorder called idiopathic CD4⁺ T-cell lymphocytopenia (ICL). ICL is a rare disorder of the immune system, defined by persistent CD4⁺ lymphopenia in the absence AIDS or any other cause of immunodeficiency. It can present with severe opportunistic infections (OIs), most commonly disseminated cryptococcosis, mycobacteria, candida, and varicella [1, 2].

Interestingly, though overlap exists among OIs in ICL and AIDS patients, the OIs with the highest incidence in ICL patients are cryptococcal and mycobacterial compared to pneumocystis and candidiasis in AIDS patients [2]. Although the etiology of ICL is unknown, proposed mechanisms include CD4⁺ lymphocyte loss due to increased activation and turnover or accelerated CD4⁺ lymphocyte apoptosis [3]. The disease can remain clinically silent, but most commonly manifests in adulthood upon acquiring an OI [4].

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Figure 1: Ulcerated 2mm papule with central crusting and surrounding erythema on the patient's forearm (arrow).

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