A rare disorder in an orphan disease: Kikuchi–Fujimoto disease in a young-adult patient with sickle cell anemia

Elisa Vencato,[1] Riccardo Manfredi,[2] Alberto Zamò,[2] Marco Chilosi,[2] Serena Beccari,[2] and Lucia De Franceschi[1]*

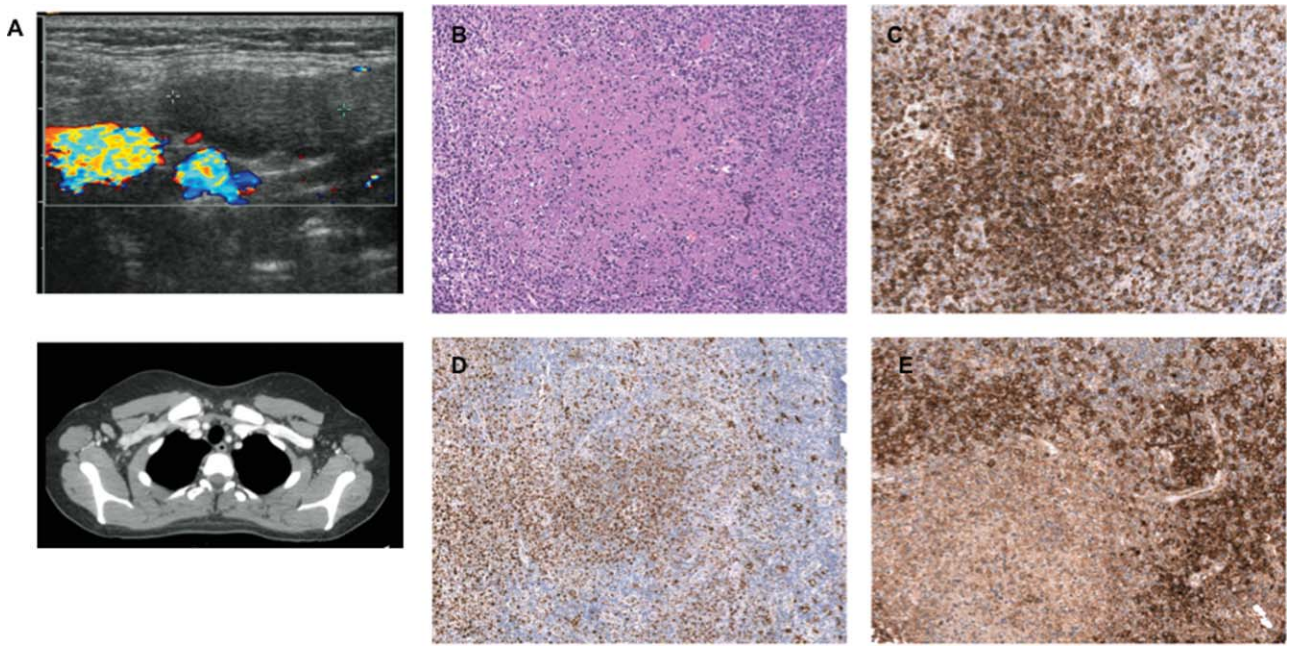

Image 1. (A) Upper panel: Color Doppler sonography of the neck. The sonographic scan shows multiple enlarged laterocervical lymph nodes. Lower panel: Contrast enhanced Computed Tomography (CT) of the chest. The CT scan shows enlarged lymph node in the right axilla. (B–E) Lymph node histological and immunohistochemical findings. (B) Typical KFD lesion with a central area of necrosis along with apoptotic bodies and karyorrhectic nuclear debris (HE, 200× magnification); (C) T-lymphoid cells with cytotoxic CD8+ phenotype surrounding necrotic area (200× magnification); (D) accumulation of CD68+ histiocytes (100× magnification); and (E) CD123+ plasmocytoid dendritic cells at the periphery of the lesions (200× magnification).

Kikuchi–Fujimoto disease (KFD) is a rare benign lymphadenopathy, also known as histiocytic necrotizing lymphadenitis [1]. KFD is a self-limited disease that predominantly affects young women and is generally associated with autoimmune pathologies [1]. In October 2013, a 22-year-old North-African female with sickle cell disease (SCD) under hydroxyurea treatment (HU; 20 mg/kg/d) presented with achy, enlarged, and tender left-side lateral cervical lymph nodes without fever and weight loss. The patient also described intense neck pain with head movements and ear-pain. Complete blood count (CBC) showed mild anemia (Hb 10.2 g/dL), normal leukocyte count (6,550/μL) with a relative reduction of total lymphocytes (2,560/μL). Immunophenotyping showed decreased T-lymphocyte count (1,515/μL), increased B-lymphocyte count (816/μL) with a decrease in the T4/T8 ratio (1.31). Erythrocyte sedimentation rate, C-reactive protein, beta-2 microglobulin levels liver, and kidney functional tests were normal. Serologic tests for Epstein–Barr and Citomegaloviruses, HIV, *Toxoplasma gondii, Francisella tularensis, Bartonella henselae, Streptococcus pyogenes, and* quantiferon-test were all negative. The antinuclear antibodies (ANA) and anti extractable nuclear antigens antibody (ENA) were negative. Ultrasonography of the neck revealed multiple enlarged laterocervical lymph nodes (Image 1A, upper panel) and computed tomograph demonstrated the presence of enlarged lymph nodes in the right axilla (Image 1A, lower panel).

[1]Department of Medicine, University of Verona and AOUI-Verona, Verona, Italy; [2]Department of Pathology and Diagnostics, University of Verona and AOUI-Verona, Verona, Italy

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***Correspondence to:** Lucia De Franceschi, M.D.; Department of Medicine, Section of Internal Medicine, University of Verona and AOUI-Verona, Policlinico GB Rossi, P.le L. Scuro, 10, 37134 Verona, Italy. Tel.: +390458124918. Fax: +390458027473. E-mail: lucia.defranceschi@univr.it.
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The biopsy of a lateral cervical lymph node revealed a partial effacement of the architecture, with patchy lesions showing central necrosis along with apoptotic bodies and karyorrhectic nuclear debris; plasmocytoid dendritic cells, activated T-lymphoid cells with cytotoxic CD8+ phenotype and hystiocites surrounded the necrotic area (Image 1B–E). The histological findings, together with the laboratory data and clinical presentation were suggestive of KFD [1]. Glucocorticoids are the most commonly used therapeutic agent for KFD [1], but in SCD patients carry the risk of worsening disease complications [2,3]. Thus, HU was halted and steroid treatment (prednisone 15 mg/die) was initiated in combination with erythrocytapheresis. We observed a rapid reduction of painful neck symptoms and a gradual reduction in size within 4 months of steroid treatment. Two young-adult female patients with SCD and KFD have been previously reported. One was under HU treatment at the moment of KFD diagnosis and both of them developed thyroid disease thereafter [4]. In summary, this is the first reported case of KFD in a SCD patient in the absence of autoimmune disease(s) treated with low dose glucocorticoids and erythrocytapheresis.

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