Vol.5 No.1

Euro Clinical Pediatrics 2020: A frequent sign, an infrequent pathology: Kikuchi-Fujimoto Disease in a Clinical Case Mariana Casullo- Castro Rendon Regional Hospital

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CASE DESCRIPTION:

Healthy twelve-year old female patient, with a clinical history of cervical adenopathy at 3 years of age. This was assumed to be an inflammatory adenitis with focal necrosis of probable viral origin. Currently, she seeks primary care due to mobile, non-tender lateral cervical adenomegalies, with a twenty-day evolution, which are associated with leukopenia without neutropenia. She refers pain to the touch (intermittent fever, myalgia and livedo reticularis in lower limbs). Not responsive to antibiotics treatment. In subsequent controls, plateletpenia and a slight anemia are detected. Neck-ultrasound shows hypervascular adenomegalies, with a tendency to confluence and a loss of hilum. EBV, CMV, and other serologies come back negative, as do rheumatological antibodies and PPD. A smear and a bone marrow aspiration eliminate the possibility of an oncological disease. The lymph node biopsy determines necrotizing histiocytic lymphadenitis. Histological comparison is performed with the previous sample, and found to match. Kikuchi-Fujimoto is diagnosed. Many studies in the literature have tried to elucidate the cause of KFD, but at present it remains unclear. The 2 most common theories explored in the literature are infectious and autoimmune. The clinical picture of KFD is similar to that of viral infection, and, like viral infection, the disease does not respond to antibiotics. Moreover, histopathologic features (ie, paracortical expansion, immunoblastic proliferation, necrosis in paracortex, predominance of T cells, and circulating atypical/reactive lymphocytes in the peripheral blood) are similar to those seen in viral infections.¹⁻³ Numerous viruses have been proposed as possible etiologic agents of KFD, including Epstein-Barr virus; herpes simplex virus; varicella zoster virus; human herpesviruses 6, 7, and 8; parvovirus B19; paramyxovirus; parainfluenza virus; rubella; cytomegalovirus; hepatitis B virus; human immunodeficiency virus; human T-lymphotropic virus type 1; and dengue virus. However, no study in the literature has definitively proven a causal relationship between a virus and KFD or identified viral particles ultrastructurally. Other infectious agents studied include Brucella, Bartonella heselae, Yersiniaenterocolitica, Toxoplasmagondii, Entamoeba histolytica, and Mycobacterium szulgai. It has been postulated that KFD represents an exuberant T-cell-mediated immune response to a variety of antigens in genetically susceptible people. Compared with the general population, patients with KFD more frequently have particular human leukocyte antigen (HLA) class II alleles, specifically HLA-DPA1 and HLA-DPB1. These alleles are more prevalent in Asians and are extremely rare or absent in whites, which may account for the more common occurrence of this entity among Asian people.

Kikuchi-Fujimoto disease has also been described in association with a number of systemic diseases, most commonly autoimmune conditions such as systemic lupus erythematosus (SLE), Wegener granulomatosis, Sjögren syndrome, Graves disease, Still disease, etc. The cytoplasm of stimulated lymphocytes and histiocytes of KFD patients contains tubular reticular structures that, by electron microscopy, bear similarity to structures that have been described in the endothelial cells and lymphocytes of patients with SLE and other autoimmune disorders. Imamura et al hypothesized that KFD may represent a self-limited, SLE-like autoimmune disorder triggered by viruses and other infectious agents. However, in contrast to SLE patients, serologic tests (such as rheumatoid factor, antinuclear antibodies, and anti-double-strand DNA antibodies) have been consistently negative in KFD patients. Despite negative serology in KFD, there is a degree of clinical and morphologic overlap between KFD and SLE that requires particular consideration. In a study by Dumas et al that included 91 KFD patients, 11 patients (12%) had a history of SLE. These cases likely represent lupus lymphadenitis, as the 2 disorders are histologically indistinguishable in some cases

Discussion and conclusion:

ECKF can affect patients of all ages, including very young children. An infectious etiology has not been proven, and it is considered to be a hyper-immune reaction. Evolution is often benign, with a limited course, which resolves within two to six months. Relapses, such as the one observed in this patient, are infrequent and have been observed in only 4% of the cases. The adenopathy must be extracted in its entirety for diagnostic purposes, since partial biopsies or punctures can render unreliable results. Long-term follow-up care of the patient is crucial