Vogt-Koyanagi-Harada Syndrome and polyarthritis as a rare clinical manifestation

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Dear Editor,

Vogt-Koyanagi-Harada Syndrome (VKHS) is a rare autoimmune inflammatory multisystem condition that mainly affects skin and the ocular, auditory and central nervous systems[1]. We report the first case of a patient with VKHS diagnosis and an erosive seronegative polyarthritis in Portugal.

This case report relates to a Caucasian 44-year-old woman with a twenty-year history of decreased visual acuity, blurred vision and cutaneous findings (alopecia and vitiligo). Ophthalmological examination revealed non-infectious chronic ocular inflammation, sign of recurrent anterior uveitis. Thus, a diagnosis of an incomplete VKHS, according to the revised diagnostic criteria of the VKH Committee was performed about ten years ago. The patient was referred by ophthalmology to the rheumatology consultation due to these ocular inflammatory changes associated with inflammatory arthralgias affecting hands, knees and feet and prolonged early morning stiffness with twenty years' duration. There was no history of inflammatory back pain, fever, skin rash or ulcers, gastrointestinal or genitourinary manifestations. The patient had no known personal or family history of rheumatic diseases or psoriasis. The musculoskeletal examination revealed symmetrical polyarthritis with involvement of four proximal interphalangeal of hands and two metacarpophalangeal joints and knees pain on palpation with patellar tap test positive bilaterally. Lumbar region and sacroiliac joint examinations were normal. On investigation, hemogram, platelet, hepatic and renal function, urinalyses, erythrocyte sedimentation rate and C-reactive protein within normal ranges (under therapy with nonsteroidal anti-inflammatory drug (NSAID)). Rheumatoid Factor (RF), anti–citrullinated protein antibody (anti-CCP), antinuclear antibody (ANA) and extractable nuclear antigens (ENA) were negative. Her serological tests were positive for anti-toxoplasma immunoglobulin IgG, anti-Rubella IgG and anti-cytomegalovirus IgG. The X-rays revealed small erosions of fourth left proximal interphalangeal and first right metacarpophalangeal joint in the hands and no erosions on the feet (Figures 1 and 2). The ultrasonography showed a synovial hypertrophy of three right proximal interphalangeal joints, without power Doppler signal. Based on the clinical course and imaging findings a diagnosis of erosive seronegative symmetrical polyarthritis was made and treatment with NSAID, short course of prednisolone 7.5 mg/day and methotrexate 10 mg/week was started with arthritis resolution.

In short, the presented case described a patient with a VKHS and a seronegative symmetrical polyarthritis with prolonged morning stiffness. There were erosions in two joints of hands and synovial hypertrophy of proximal interphalangeal joints. Although corticoid is commonly used in the treatment of VKHS, methotrexate is also added because a diagnosis of seronegative polyarthritis is assumed.

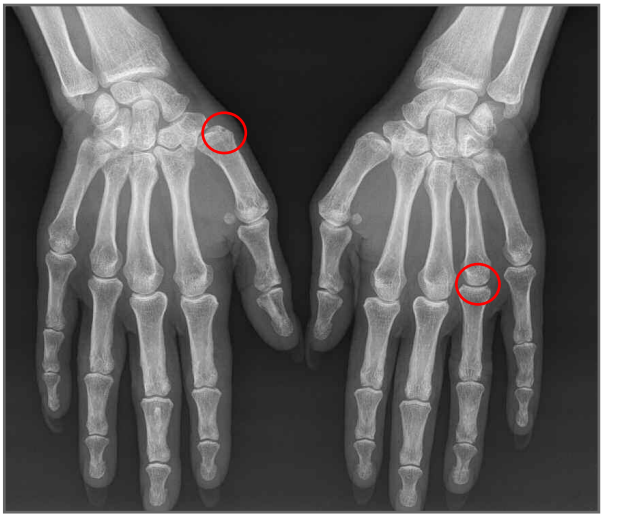

FIGURE 1. Hands X-ray. Small erosions of fourth left proximal interphalangeal and first right metacarpophalangeal joint.

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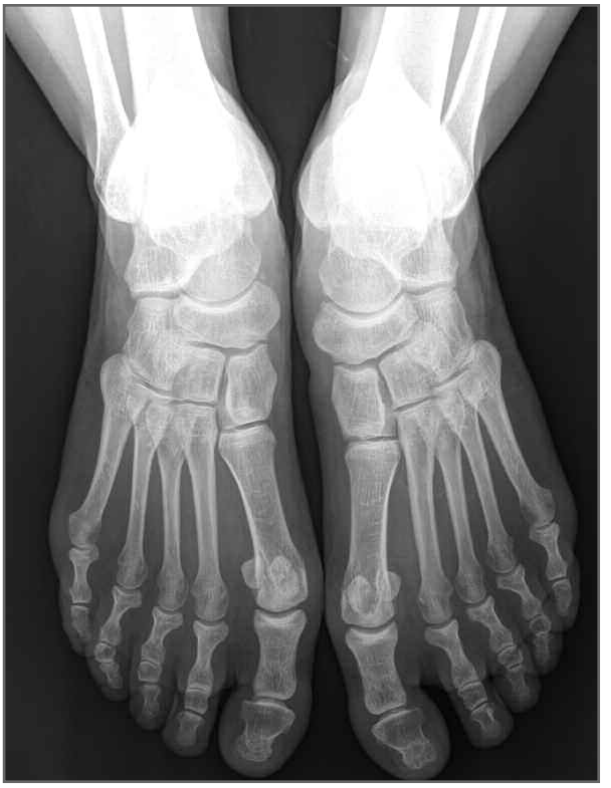

FIGURE 2. Feet X-ray. No erosions.

This case supports the hypothesis that polyarthritis is part of the clinical spectrum of the VKHS. Although the inflammatory aetiology of VKHS is not completely unclear, it is proposed that cytotoxic T cells target the surface of melanocytes of various systems in genetically susceptible individuals[1]. On the other hand, RA is a chronic systemic inflammatory disorder with etiology not completely clarified which primarily affects the joints. Both VKHS and RA are systemic inflammatory diseases with common eye involvement (as uveitis or keratoconjunctivitis sicca and scleritis/episcleritis, respectively)[2]. Hence, these two conditions probably share an autoimmune inflammatory origin that involves an imbalance of Th1/Th2 and Th17/Treg cells with dominance of inflammatory cytokines. Moreover, recent research established that both diseases share some features of the genetic susceptibility such as human leukocyte antigen (HLA)-DR4, HLA-DR1, CTLA-4 and STAT4[3-5]. Despite there is a potential link between VKHS and polyarthritis, further research is needed to explore and confirm this association. This case highlights that a high index of clinical suspicion and a multidisciplinary approach with ophthalmologists and rheumatologists are needed to establish an early diagnosis of arthritis in these patients and consecutively, prevent structural damage.

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