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Case report

Rhupus syndrome. A rare combination☆

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ABSTRACT

Rhupus syndrome is a rare combination of rheumatoid arthritis and systemic lupus erythematosus, and is characterized by the presence of erosive arthritis together with symptoms and signs of systemic lupus erythematosus. Among its complications, is the presence of rheumatoid nodules, and neurological and renal involvement that further complicates its prognosis, thus significantly reducing the perception of health-related quality of life in patients who suffer from it. The case is presented of a female patient diagnosed with lupus erythematosus, who during the course of the disease, developed clinical and humoral signs that led to the diagnosis of syndrome Rhupus syndrome. This is believed to of relevance to the knowledge of the medical community.

Síndrome de Rhupus. Una superposición infrecuente

Palabras clave:
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Lupus eritematoso sistémico
Síndrome de Rhupus

RESUMEN

El síndrome de Rhupus es una rara superposición de artritis reumatoide y lupus eritematoso sistémico, que se caracteriza por la presencia de una poliartritis erosiva asociada a síntomas y signos de lupus eritematoso sistémico. Entre sus complicaciones destaca la presencia de nódulos reumatoideos, afectación neurológica y renal que complican más aún la evolución

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del caso, disminuyendo considerablemente la percepción de la calidad de vida relacionada con la salud de los pacientes que lo padecen. Se presenta el caso de una paciente mujer con diagnóstico de lupus eritematoso, que desarrolla en el curso de la enfermedad manifestaciones clínicas y humorales que permiten llegar al diagnóstico de un síndrome de Rhupus, lo cual consideramos sea de importancia para el conocimiento de la comunidad médica.

Introduction

Rheumatic diseases (RD) include a group of about 250 conditions that primarily affect the bones, muscles and joints, being characterized clinically by the presence of pain, inflammation, stiffness, deformity and different degrees of disability, that cause a decreased perception of the health related quality of life (HRQoL).[1,2]

Despite the fact that there are well-established diagnostic criteria, up to 25% of patients with RD, with systemic symptoms, cannot be clearly diagnosed. They are patients who share clinical and pathological characteristics of systemic inflammation without meeting the criteria for a specific disease.[3] One of the elements that brings more difficulty in this regard is the overlap of RD in a single entity. Multiple overlaps are described, that even come to be considered as an independent entity, as it is the case of the mixed connective tissue disease.[4,5]

Other overlaps are reported, among them, one of the most discussed and infrequent is the one that combines clinical and laboratory elements of rheumatoid arthritis (RA) and systemic lupus erythematosus (SLE). The first reports on this disease date back to the year 1960 when the first clinical observations that helped to identify this entity were made. However, it was not until the year 1971 when the term Rhupus was used for the first time to refer to this condition.[4–6]

Overlap syndromes are considered a rare phenomenon. Rhupus syndrome (RhS) has been estimated at between 0.01% and 2% of patients with RD. There is a variant of overlap between juvenile idiopathic arthritis and SLE, which is described as a rare clinical condition in children.[7,8]

RhS is defined as a deforming and erosive symmetric polyarthritis accompanied by symptoms of SLE and the presence of antibodies of high diagnostic specificity, such as anti-double stranded DNA, anti-Smith and anti-cyclic citrullinated peptide (anti CCP) antibodies.[9] Renal involvement is characteristic of this syndrome; several authors have encountered findings of type IV lupus nephritis in patients diagnosed with RhS,[9,10] which together with the neurological manifestations causes a decrease in the perception of HRQoL.[11–14]

That is why, taking into account the infrequent occurrence of this condition, its repercussion on the osteo-muscle-articular system and the renal complications that it causes, the impact on the perception of the HRQoL in the patients who suffer from it and the scarcity of reports about the disease in Ecuador, it was decided to carry out this work with the purpose to let the medical community know the clinical and humoral manifestations of RhS.

Case presentation

A 47-year-old female patient with a diagnosis of SLE for 6 years, currently treated with 50 mg of azathioprine daily, 7.5 mg of prednisone daily, 250 mg of chloroquine daily and 100 mg of acetylsalicylic acid daily. At the time of the onset of the disease she met 5 diagnostic criteria for SLE (malar rash, oral ulcers, positive antinuclear antibodies [ANAs], complement consumption and thrombocytopenia). During the 6 years of evolution of the disease she presented periods of exacerbation with predominance of joint manifestations and constitutional symptoms, given by the presence of asthenia, anorexia, weight loss and low-grade fever in the evening, which resolved with the increase in the dose of steroids, which in some occasions reached up to 20 mg daily.

On this occasion she attends the consultation reporting that for approximately 5 months she has had a polyarticular additive inflammatory clinical picture that involves primarily the small joints of the hands and feet, as well as the left elbow, the right knee and both temporomandibular joints. It is accompanied by morning stiffness for about 90 min and exacerbation of the constitutional symptoms given by asthenia, anorexia and low-grade fever in the evening. The patient states that she has had these symptoms in other occasions, but of less intensity and duration, which disappear when the dose of steroids is increased. In addition she has right frontal-occipital headache of moderate intensity, photosensitivity, sleep disorders, irritability and affective lability.

On physical examination are found as significant data the presence of slight malar rash, alopecia of 2 cm in diameter in the right frontal-parietal region, limited mobility of the cervical segment and limitation of the flexion/extension of both wrists (30°). The left elbow shows inflammatory signs given by pain both spontaneous and induced by mobilization, heat and increase of volume; at the level of the hands can be observed atrophy of the interosseous muscles, bilateral swelling of the second and third metacarpophalangeal joints and the proximal interphalangeal joints (PIPs), hyperflexion deformity in distal interphalangeal joints (DIPs) and bilateral ulnar deviation of the fingers (Fig. 1). It can be observed the presence of joint effusion in the right knee, with increased local temperature, pain at digital pressure in the articular interline with predominance of the internal compartment, with pain at digital pressure on the surface of the internal femoral condyle; in addition, pain at digital pressure of the right anserine bursa. In the feet is found thrusting of the 2nd finger over the 3rd finger of the right foot, bilateral hallux valgus and bilateral positive *pullison* maneuver.