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Case-based Review

# Multifocal osteonecrosis in systemic lupus erythematosus: Two case reports and literature review

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## Abstract

Multifocal osteonecrosis (MFO) is an uncommon and disabling condition. A few cases have been described in association of systemic lupus erythematosus (SLE). We describe 2 clinical cases of patients with SLE along with MFO and also perform review of the literature.

**Keywords:** Osteonecrosis, systemic lupus erythematosus, corticosteroids

## Introduction

We present 2 cases of multifocal osteonecrosis (MFO) in 2 patients with systemic lupus erythematosus (SLE), involving 9 anatomical areas revealed by magnetic resonance imaging (MRI) (Ethics Committee Approval: CSAPG-UT 07/2019) (Table 1).

## Case Presentation

The clinical cases are as follows: I) a 52-year-old Caucasian woman having 11 years of SLE disease duration at the first osteonecrosis lesion evidence (knee). The main SLE involved systems included affected joints and skin, presence of Raynaud's phenomenon, and antinuclear antibodies (ANA) and anti-double-stranded DNA antibody (anti-dsDNA) antibodies positivity; moreover, she had received up to 20 mg of prednisone (Figure 1 a-e; MRI images of osteonecrotic areas). II) A 44-year-old Caucasian woman having 3 years of SLE disease duration at the first osteonecrosis lesion evidence (wrist). The main SLE involved systems included joints, skin, constitutional symptoms, hematological, kidney, serositis, and ANA, anti-dsDNA, and anti-RNP antibodies positivity; she had received up to 40 mg of prednisone throughout life (Figure 1f; MRI image of osteonecrotic areas).

## Discussion

MFO is defined by the presence of osteonecrotic lesions in at least 3 locations (1), corticosteroid exposure being a common denominator in most cases (2). A total of 26 MFO cases have been described in the literature affecting patients with SLE (1, 3, 4). Therefore, MFO is a very uncommon condition in SLE, although some authors have suggested that it may cause 38% of all MFOs (2). Knee joint is the most frequently affected site. The lapse between the SLE diagnosis and the presence of the first osteonecrotic lesion may be 6 years on average. The number of affected sites in different MFOs varies between 4 and 28 anatomical sites. ANA and anti-DNA antibodies are present in most cases. The factors that are most related to the presence of osteonecrosis in SLE include use of corticosteroids, disease activity, and cytotoxic drugs (5).

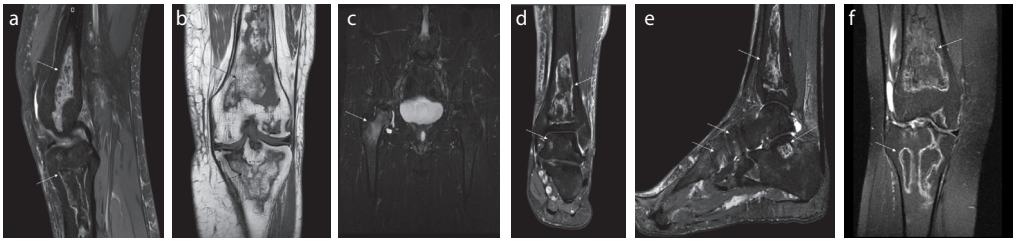


**Figure 1. a-f.** Multiple osteonecrotic areas (arrows) suggested by nuclear magnetic resonance imaging: Longitudinal short-TI inversion recovery (STIR) (a) and coronal T1-weighted magnetic resonance image showing proximal tibia and distal femur section (b); coronal STIR magnetic resonance image showing proximal femur section (c); coronal STIR magnetic resonance image showing distal tibia and talus section (d); longitudinal STIR magnetic resonance image showing distal tibia, calcaneus, and first cuneiform bone section (e); and coronal STIR magnetic resonance image showing proximal tibia and distal femur section (f).

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**Table 1.** Clinical and anatomical description of 2 patients with systemic lupus erythematosus who presented multifocal osteonecrosis.

| Age | Sex | Ethnicity | SLE disease duration | Clinical involvement | Immunological profile | Antiphospholipid antibodies | SLE-related treatments | Maximum steroids dosage (prednisone) | Number and involved areas of multifocal osteonecrosis | Outcome |
|---|---|---|---|---|---|---|---|---|---|---|
| 52 | Female | Caucasian | 11 | Arthritis<br>Malar rash<br>Photosensitivity<br>Oral and ocular dryness<br>Raynaud phenomenon | ANA<br>dsDNA | Negative | HCQ | 20 mg | 9 areas:<br>Bilateral proximal femur<br>Bilateral distal femur<br>Bilateral proximal tibia<br>Left medial and intermediate cuneiform bones<br>Left calcaneus<br>Left navicular (foot)<br>Left fifth metatarsal<br>Left proximal humerus | Bilateral hip and left knee prosthetic replacement |
| 44 | Female | Caucasian | 3 | Arthritis<br>Malar rash<br>Photosensitivity<br>Fever<br>Lymphopenia<br>Low platelet count<br>Pleuritis<br>Pericarditis<br>Proteinuria | ANA<br>dsDNA<br>RNP<br>Histona | Negative | HCQ<br>AZA* | 40 mg | 9 areas: Bilateral proximal tibia<br>Bilateral distal femur<br>Right left navicular femur(hand)<br>Left semilunar and navicular (hand)<br>Bilateral proximal femur | Bilateral hip and left knee prosthetic replacement |

*Treatment discontinuation due to disease remission.
SLE: systemic lupus erythematosus; g: grams; ANA: antinuclear antibodies; DNA: anti-dsDNA antibodies; RNP: ribonucleoprotein; HCQ: hydroxychloroquine; AZA: azathioprine; mg: milligrams.

These are the factors that reproduce in different published cases of MFO, in addition to stating the presence of antiphospholipid antibodies in some of them (1, 3, 6). The patients' informed consents were obtained.

## Main Points

- Systemic lupus erythematosus may present multifocal osteonecrosis through follow-up.
- Rheumatologists may suspect the presence of multifocal osteonecrosis in asymptomatic patients with systemic lupus erythematosus.
- Steroid dosages may reduce the potential presence of multifocal osteonecrosis in patients with systemic lupus erythematosus.

**Informed Consent:** Informed consent was obtained from the patients.

**Peer-review:** Externally peer-reviewed.

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