Radiology Case Reports 13 (2018) 915–919

Available online at www.sciencedirect.com

ScienceDirect

journal homepage: www.elsevier.com/locate/radcr

Case Report

Magnetic resonance enterography appraisal of lupus enteritis: A case report

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ARTICLE INFO

Article history:
Received 27 March 2018
Revised 8 June 2018
Accepted 12 June 2018

Keywords:
Magnetic resonance enterography
Lupus enteritis

ABSTRACT

Systemic lupus erythematosus (SLE) is a chronic autoimmune disease with a multisystemic involvement. Usually, radiological imaging does not play a central role in evaluating SLE patients, although it may be helpful in assessing complications, allowing a more accurate evaluation of the patient. Lupus enteritis is one of the most common and potentially lethal manifestations of the gastrointestinal involvement of SLE. Among the imaging modalities, computed tomography scan is now considered the gold standard in evaluating lupus enteritis, although it is impaired by the radiation exposure. On the other hand, during the last decade magnetic resonance enterography has achieved a remarkable importance in evaluating small bowel lesions in patients affected by Crohn's disease. We describe the first case report of lupus enteritis evaluated with magnetic resonance enterography, putting forward the proposal of a reliable and radiation-free alternative to computed tomography scan in evaluating the intestinal involvement of SLE.

Introduction

Systemic lupus erythematosus (SLE) is a chronic multisystemic autoimmune disease whose specific etiology still remains unknown [1,2].

A genetic predisposition and some environmental risk factors contribute to its onset, leading to an altered immune response consisting in hyperactivation of T and B lymphocytes, loss of self-tolerance, and formation of circulating pathogenic immune complexes, with their consequent deposition and damage of several organs [1,2].

The overall incidence rates for SLE are approximately 0.3-23.7 per 100,000 person-years, with a prevalence that range from 6.5 to 178.0 per 100,000 and a female–male ratio close to 9:1 [2,3].

Although the diagnosis and the evaluation of the disease as a whole are strictly clinical, the assessment and the follow-up of some complications may require the usefulness of radiological imaging.

In particular, the gastrointestinal involvement of SLE is a potentially severe complication of SLE [4], with an incidence that range from 5.4% to 40% of the patients [5,6]; among its possible clinical manifestations, one of the most common

☆ Competing Interests: None.

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https://doi.org/10.1016/j.radcr.2018.06.008

is lupus enteritis, an immunocomplex-mediated vascular inflammation that may lead to the necrosis of the vessel walls [5,7].

According to the definition given by the British Isles Lupus Assessment Group disease activity index, lupus enteritis is intended as a "gastrointestinal SLE involvement as either vasculitis or inflammation of the small bowel, with supportive imaging and/or biopsy findings" [8]. However, in literature, lupus enteritis and lupus vasculitis are often used as synonyms, together with other denominations, such as mesenteric arteritis, lupus arteritis, gastrointestinal vasculitis, intra-abdominal vasculitis, and acute gastrointestinal syndrome [5,9,10].

Up to now, all the different imaging modalities have not shown pathognomonic signs related to lupus enteritis, including computed tomography (CT) scan, that is considered the gold standard investigation in spite of the radiation exposure.

Magnetic resonance enterography (MRE) is a radiation-safe, full comprehensive examination usually indicated for patients affected by Crohn's disease (CD).

However, considering the increasingly importance that this technique has achieved during the last years in evaluating small bowel lesions, it is possible to consider new frontiers of its performing.

To our knowledge, we describe the first case report of gastrointestinal involvement of SLE evaluated with MRE.

Case report

We describe the case of a 22-year-old woman affected by SLE who had been hospitalized twice, in 2 different hospitals, due to gastrointestinal symptoms.

The first time, an abdominal x-ray plain radiograph and a CT scan were obtained, showing some gas–fluid levels within the ileal loops, whose walls were also thickened and with a layered aspect; some centimetric lymph nodes were also visible in the mesenteric fat, and perihepatic and perisplenic fluid collections were seen.

A biopsy through a colonoscopic exam was also performed, which showed mucosal ulcerative lesions in the terminal ileum with cellular infiltration and hemorrhage foci within the underlying layers of the intestinal wall, allowing the diagnosis of lupus enteritis.

Moreover, a US examination of both kidneys and an ultrasound-guided biopsy of the lower pole of the left kidney were already performed, demonstrating a renal histology of class IV lupus nephritis.

The patient was discharged after the prescription of steroids and immunosuppressive therapy.

However, the immunosuppressive therapy was later suspended due to the onset of a marked neutropenia.

After 10 months from the last hospitalization, the patient came to the Emergency Room of our hospital due to the recrudescence of the abdominal symptoms and the occurrence of vasculitic urticaria with angioedema of the right eye and the superior lip.

Laboratory tests showed active renal disease, with increased proteinuria (3040, 70 mg/24 h), low complement fraction C3 (61, 9 mg/dL), low C4 (5.29 mg/dL), increased PCR (31, 54 mg/L), high velocità di eritrosedimentazione (VES) value (40 mm/h), positive elevated anti-ds-DNA antibodies (123, 60 IU/mL), positive antinuclear antibody at 1:1600, positive anti-Ro antibodies, and a normal lymphocyte count with lower CD4+ and/or CD8+ ratio.

In order to assess the current status of the intestinal involvement and in accordance with the clinicians, it was decided to perform an MRE, with the principal aim of sparing the patient another amount of radiations.

MRE requires the oral administration of approximately 1500 mL of polyethylene glycol-water solution, starting 45 minutes before the beginning of the exam.

After the patient was placed in supine position inside the scanner, coronal thick-section T2-weighted rapid acquisition with relaxation enhancement (RARE) acquisition, axial and coronal T2-weighted true fast imaging with steady-state precession (repetition time/echo time: 4.20/2.10 ms, flip angle (FA): 60°), and half-Fourier acquisition single-shot turbo spin echo (repetition time/echo time: ∞/80 ms) with and without fat suppression were performed, together with diffusion-weighted imaging (DWI) sequences, obtained on the axial plane using a diffusion factor b fixed at 0, 400, and 800 s/mm^2.

Coronal precontrast ultrafast 3D T1-weighted gradient-echo fat-suppressed and ultrafast axial 3D T1-weighted gradient-echo fat-suppressed images obtained after injection of gadoterate meglumine (Dotarem) at a dose of 0.2 mL/kg body weight were acquired at 30, 60, and 180 seconds, followed by a bolus of 30 mL of normal saline.

The exam allowed to detect a mild thickening (5 mm) of several ileal loops, whose total extension, measured with digital calipers from the ileocecal valve, amounted to 38 cm.

Moreover, the "thumb printing sign," usually related to ischemic condition, was clearly detectable on T2-weighted thick-section RARE images. A moderate amount of free fluid was also seen within the abdominal cavity (Fig. 1).

Diffusion-weighted and apparent diffusion coefficient (ADC) calculation did not show water restriction, whereas contrast-enhanced sequences demonstrated a mild enhancement of the thickened small bowel walls (Fig. 2).

The patient was treated with steroids (intravenous administration of methylprednisolone, 1 g/day for 3 days) and monoclonal antibodies (intravenous administration of rituximab, 1g/day).

After the relief of the abdominal and cutaneous symptoms and the prescription of the steroid therapy, the patient was discharged.

Discussion

Although several manifestations of the gastrointestinal tract involvement can be recognized in SLE patients (eg, protein-losing enteropathy, intestinal pseudo-obstruction, eosinophilic enteritis, etc.) [7], lupus enteritis remains one of the most common, affecting up to 53% of the patients presenting abdominal pain [4,11].

Inflammatory enteritis is consequent to the deposition of circulant pathologic immunocomplex and thrombosis of the intestinal vessels [12]; its prevalence ranges from 0.2% to 53%