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Abdominal Manifestations of Systemic Lupus Erythematosus: Spectrum of Imaging Findings

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Abstract

Systemic lupus erythematosus is an immune-mediated syndrome in which the immune response is to non−organ-specific antigens, and virtually every organ in the abdominal cavity may become involved. Only renal involvement forms part of the diagnostic criteria, however, a combination of typically nonspecific findings, including peritoneal surface, enteric, renal, renal tract, pancreatic, adrenal, hepatobiliary, and splenic manifestations, should be looked for in patients with known lupus or other connective tissue disease who are undergoing abdominal imaging and may suggest the diagnosis in patients presenting with an acute abdomen. Our work presents the spectrum of imaging findings of abdominal manifestations of systemic lupus erythematosus.

Abrégé

Le lupus érythémateux disséminé est une maladie auto-immune présentant une réponse immunitaire non propre à un organe. En effet, tous les organes de la cavité abdominale peuvent être touchés. Si l'atteinte rénale constitue un des critères de diagnostic, une combinaison de résultats habituellement aspécifiques, comme les manifestations péritonéales, entériques, rénales, du tractus rénal, pancréatiques, surrénaliennes, hépatobiliaires et spléniques, doit être recherchée chez les patients souffrant de lupus ou d'une autre maladie du tissu conjonctif qui doivent passer un examen d'imagerie abdominale. Ces signes peuvent également suggérer un diagnostic chez les patients avec un abdomen aigu. Nous présentons le spectre complet des résultats d'imagerie des manifestations abdominales du lupus érythémateux disséminé.

Key Words: Abdominal imaging; Computed tomography; Lupus enteritis; Lupus vasculitis; Systemic lupus erythematosus

Systemic lupus erythematosus (SLE) is a disease of unknown etiology in which tissues and cells are damaged by pathogenic auto-antibodies and immune complexes. Up to 90% of affected patients are female. The clinical course is characterized by remissions and exacerbations, with mucocutaneous, cardiorespiratory, renal, and neurologic manifestations. Despite established diagnostic criteria (Table 1) [1], many patients with SLE also demonstrate features of other connective tissue diseases, such as dermatomyositis or systemic sclerosis.

Abdominal manifestations may result from primary involvement with SLE or from complications of therapy and are well documented in the rheumatology literature [2−5]. Serositis and intestinal vasculitis can present with severe abdominal pain and may masquerade as an acute surgical abdomen. Small and large bowel ischemia may result from vasculitis or vascular occlusion associated with the antiphospholipid syndrome. Renal involvement is common and forms part of the diagnostic criteria (Table 1). The liver, spleen, pancreas, adrenals, and genitourinary tract may also become involved [2−4].

While the imaging appearances are typically nonspecific, retrospective reviews suggest a diagnostic role for computed tomography (CT) in patients with lupus enteritis [6−8]. Peritoneal surface, renal, renal tract, adrenal, hepatobiliary, pancreatic, and splenic manifestations should be looked for in patients with known lupus or other connective tissue disease undergoing abdominal imaging and may suggest the diagnosis in a patient presenting with an acute abdomen.

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Table 1
The 1982 Revised Criteria for Classification of Systemic Lupus Erythematosus*

Criterion	Definition
1. Malar rash	Fixed erythema, flat or raised, over the malar eminences, tending to spare the nasolabial folds
2. Discoid rash	Erythematous raised patches with adherent keratotic scaling and follicular plugging; atrophic scarring may occur in older lesions
3. Photosensitivity	Skin rash as a result of unusual reaction to sunlight, by patient history or physician observation
4. Oral ulcers	Oral or nasopharyngeal ulceration, usually painless, observed by physician
5. Arthritis	Nonerosive arthritis involving 2 or more peripheral joints, characterized by tenderness, swelling, or effusion
6. Serositis	a) Pleuritis: convincing history of pleuritic pain rubbing heard by a physician or evidence of pleural effusion. or b) Pericarditis: documented by electrocardiogram or rub or evidence of pericardial effusion
7. Renal disorder	a) Persistent proteinuria greater than 0.5 g/d or more than 3 + if quantitation not performed or b) Cellular casts: may be red cell, hemoglobin, granular, tubular, or mixed
8. Neurologic disorder	a) Seizures: in the absence of offending drugs or known metabolic derangements; eg, uremia, ketoacidosis, or electrolyte imbalance or b) Psychosis: having excluded other causes, eg, drugs
9. Hematologic disorder	a) Hemolytic anemia, with reticulocytosis or b) Leukopenia : $<4.0 \times 10^9$/L on 2 or more occasions or c) Lymphopenia $<1.5 \times 10^9$/L on 2 or more occasions or d) Thrombocytopenia: $<100 \times 10^9$/L
10. Immunologic disorder	a) Positive LE cell preparation or b) Anti-DNA: antibody to native DNA in abnormal titer or c) Anti-Sm: presence of antibody to Sm nuclear antigen or d) False-positive serologic test for syphilis known to be positive for at least 6 mo and confirmed by Treponema pallidum immobilization or fluorescent treponemal antibody absorption test
11. Antinuclear antibody	An abnormal titer of antinuclear antibody by immunofluorescence or an equivalent assay at any point in time and in the absence of drugs known to be associated with ''drug-induced lupus'' syndrome

*From Ref. 1.

Our work reviews the spectrum of imaging findings of abdominal manifestations of SLE.

Serositis

Ascites occurs in approximately 10% of patients with SLE (Figure 1). Possible causes include serositis, pancreatitis, nephritis and nephrotic syndrome, constrictive pericarditis, and peritoneal inflammation secondary to vasculitis. Chylous ascites is particularly unusual (Figure 2) [9]. Ascites is an infrequent finding in patients with lupus nephritis and, when present, is more likely to be due to peritoneal serositis than hypoalbuminemia.

Serositis can develop when SLE is inactive but may also manifest as peritonitis that may mimic an acute surgical abdomen and lead to exploratory laparotomy. Low et al [10] reported the characteristics of lupus serositis on barium small bowel series as showing segments of spiculation with tethering, angulation, and obstruction. CT demonstrated ascites and asymmetric thickening of the small bowel wall. Laparotomy showed extensive patchy serosal and peritoneal plaques, but biopsies did not reveal any specific lesions [10]. Similar to vasculitis, ascites and serositis typically resolve with high-dose steroid therapy [8].

Enteric Manifestations

Gastrointestinal (GI) involvement in SLE may be easily overlooked since many GI symptoms can be attributed to complications of drug therapy, such as nonsteroidal anti-inflammatory drugs, steroids, and azathioprine. Involvement of the GI tract in SLE is almost always accompanied by evidence of active disease in other organs, although occasionally it may be the main clinical feature [2].

Mouth ulcers may occur in up to 50% of patients with SLE [3]. Mucosal ulcers usually occur in the hard palate, buccal mucosa, or the vermilion border, and may be erythematous and painless, or discoid and painful, causing dysphagia or odynophagia [4].

Dysphagia may be secondary to a lack of saliva for mastication as seen in Sjögren syndrome, infection, or esophageal dysmotility. Tertiary contractions, impaired esophageal emptying and/or esophageal dilatation may be seen on barium upper-GI series, and manometry may