BRIEF COMMUNICATION

Abdominal pain as the initial and sole clinical presenting feature of systemic lupus erythematosus

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Classically, a diagnosis of systemic lupus erythematosus (SLE) is dependent on renal, rheumatological, cutaneous and neurological target organ damage with supporting serological markers. A previously healthy 26-year-old Japanese woman whose only manifestation of otherwise occult SLE was severe abdominal pain is reported. A computed tomographic scan of the abdomen revealed thickened loops of small bowel, endoscopic findings were nonspecific and jejunal biopsy revealed a nonspecific enteritis. Laboratory studies revealed lymphopenia, hypocomplementemia, a positive antinuclear antibody, a weakly positive anti-Smith and a strongly positive anti-double stranded DNA. There was a prompt symptomatic recovery with immunosuppressive therapy. The authors’ experiences, and a review of the literature suggest that a diagnosis of SLE should be considered in young Asian women who present with significant but clinically enigmatic gastrointestinal illness.

Key Words: Abdominal pain; Systemic lupus erythematosus

A bdominal pain is a very common medical complaint, and accounts for a significant number of primary care physician visits and referrals to gastroenterologists. In one survey, 21.8% of respondents reported abdominal pain in the preceding one month, and more than 65% rated the symptoms as either moderate or severe (1). We report a case of a patient who presented with severe abdominal pain that was the initial clinical manifestation of occult systemic lupus erythematosus (SLE).

CASE PRESENTATION

A previously healthy 26-year-old Japanese woman presented with diffuse, colicky central abdominal pain, a four-week history of frequent nonbloody, soft stools, a two-week history of anorexia and fatigue and a three-day history of fever, nausea and vomiting. Movement aggravated her pain and there were no relieving factors. There was no melena, bloody diarrhea, night sweats, arthralgias, skin rashes, photosensitivity, headaches, visual disturbances, dyspnea, chest pain, Raynaud’s phenomena, sicca symptoms or changes to her bladder habits. She did not note a 2.3 kg weight loss over the past three months. Medications before presentation included oral contraceptives and occasional ibuprofen or acetaminophen. She did not smoke or consume alcohol and the family history was unremarkable. On examination, she was afebrile. Head, neck and cardiopulmonary examinations were unremarkable. The abdomen was soft but tender over the epigastrium without rebound. There was no stigmata of chronic liver disease, hepatosplenomegaly, masses or significant distention. A computed tomographic scan of the abdomen revealed thickened loops of small bowel, endoscopic findings were nonspecific and jejunal biopsy revealed a nonspecific enteritis. Laboratory studies revealed lymphopenia, hypocomplementemia, a positive antinuclear antibody, a weakly positive anti-Smith and a strongly positive anti-double stranded DNA. There was a prompt symptomatic recovery with immunosuppressive therapy. The authors’ experiences, and a review of the literature suggest that a diagnosis of SLE should be considered in young Asian women who present with significant but clinically enigmatic gastrointestinal illness.

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La douleur abdominale comme caractéristique initiale et unique de lupus érythémateux disséminé à la présentation

D’ordinaire, un diagnostic de lupus érythémateux disséminé (LED) dépend de dommages rénaux, rhumatologiques, cutanés et neurologiques d’un organe cible et de marqueurs sérologiques. On présente le cas d’une Japonaise de 27 ans auparavant en santé dont le LED autrement occulte se manifestait seulement par de graves douleurs abdominales. Une tomodensitométrie de l’abdomen a révélé des anes épaisse de l’intestin grêle, les observations endoscopiques n’étaient pas spécifiques et la biopsie jéjuna a démontré la présence d’une entérite non spécifique. Les études de laboratoire ont révélé une lymphocytopenie, une hypocomplémentémie, un anticorps antinucléaire positif, un anti-Smith légèrement positif et un brin d’ADN antitubulaire. Les symptômes ont rapidement disparu grâce à la prise d’immunosuppresseurs. D’après les expériences de l’auteur et un examen de la documentation scientifique, un diagnostic de LED devrait être envisagé chez les jeunes femmes asiatiques qui se présentent avec une maladie gastro-intestinale importante, silencieuse du point de vue clinique.
antigens (Quanta Life™ ELISA; Inova Diagnostics, USA) homogenous and speckled pattern. The extractable nuclear antinuclear antibody titre was positive at 1:160 with a mixed and 0.05g/L (normal range 0.12 to 0.36 g/L) respectively. The C4 were decreased at 0.48 g/L (normal range 0.80 to 1.80 g/L) or mesenteric and bilateral renal selections. not show any evidence of vasculitis on selective celiac, superi-
unremarkable. A mesenteric angiogram was performed that did ileum did not reveal any abnormalities. Echocardiography was
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jejenum (Figure 2) revealed a nonspecific inflammatory infil-
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the abdomen (Figure 1) confirmed these findings. Upper gas-
thickening and moderate ascites. A computed tomography of the abdomen (Figure 1) confirmed these findings. Upper gas-
testinal endoscopy was normal and the biopsy of the
jejenum (Figure 2) revealed a nonspecific inflammatory infiltrate. Colonoscopy and biopsies of the colon and terminal ileum did not reveal any abnormalities. Echocardiography was unremarkable. A mesenteric angiogram was performed that did not show any evidence of vasculitis on selective celiac, superior or mesenteric and bilateral renal selections.

Further laboratory investigations revealed that the C3 and C4 were decreased at 0.48 g/L (normal range 0.80 to 1.80 g/L) and 0.05g/L (normal range 0.12 to 0.36 g/L) respectively. The antinuclear antibody titre was positive at 1:160 with a mixed homogenous and speckled pattern. The extractable nuclear antigens (Quanta Life™ ELISA; Inova Diagnostics, USA) revealed a weakly positive anti Smith, anti- SCL 70, and anti- SSB. The anti-double stranded DNA was strongly positive at 986 IU/mL (strong positive greater than 801 IU/mL) as was the anti-SSA at 193.8 EU (strong positive greater than 80 EU) A diagnosis of SLE of the small bowel was made. Her oral con-

Figure 1) Contrast-enhanced computed tomography of the abdomen demonstrates circumferential bowel wall thickening involving a segment of jejunum (long arrow). A small amount of ascites is also present in the right paracolic gutter (short arrow).

Figure 2) Duodenal biopsy (Hematoxylin and eosin, original magnification × 252; inset original magnification × 608). The architecture is normal. There is a neutrophilic infiltrate in the superficial epithelium, predominantly in the tips of the villi, and a mild increase in lymphocytes in the deeper glands (inset). There are no infective agents or viral cytopathic effects.

thickening and moderate ascites. A computed tomography of the abdomen (Figure 1) confirmed these findings. Upper gastrointestinal endoscopy was normal and the biopsy of the jejenum (Figure 2) revealed a nonspecific inflammatory infiltrate. Colonoscopy and biopsies of the colon and terminal ileum did not reveal any abnormalities. Echocardiography was unremarkable. A mesenteric angiogram was performed that did not show any evidence of vasculitis on selective celiac, superior or mesenteric and bilateral renal selections.

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ed of azathioprine 50 mg and prednisone 20 mg daily with a prompt resolution of symptoms. Two months after her initial presentation she experienced two episodes of right sided amau-
rosis fugax. Tests for anticardiolipin antibodies and delayed Russel venom viper test (DRVVT) were negative. Carotid ultrasound, cardiac echocardiogram and magnetic resonance angiography were negative. A cetylsalicilic acid was added to the regimen.

DISCUSSION
SLE is a common disorder that affects one in 2500 people, with a female predominance (2). The classification (but not clinical diagnosis) of SLE is typically based on the 1982 revised criteria of the American Rheumatologic Association (ARA) for the classification of SLE (3), which is reported to be 96% sensitive and 96% specific. The ARA criteria require a serial or simultaneous presentation of four of 11 listed symptoms, signs or laboratory findings for a diagnosis of lupus (3). Our patient met minimal ARA diagnostic criteria of SLE but did not have any of the classic renal, dermatological (ie, malar or discoid rash), musculoskeletal (ie, polyarthritis) or neurological features that are included in the ARA criteria. Moreover, evidence of serositis (ie, pleural effusion, ascites) was apparent only on diagnostic imaging. A bdominal pain and evidence of gastrointestinal disease are not included in the ARA criteria but in our patient’s case, a diagnosis of SLE was confirmed by her lym-
phopenia, hypocomplementemia, positive antinuclear antibody and strongly positive anti-double stranded DNA, anti-SSA, as well as the weakly positive anti-Smith. Therefore, we are confident that her clinical diagnosis was SLE.

Gastrointestinal manifestations of SLE that have been reported in the literature consist of abdominal pain, nausea, vomiting and diarrhea, and are reported to occur in greater than 50% of SLE patients at some time during the course of the disease (4). A bdominal pain during a lupus flare is usually attributed to polyserositis. In cases of acute abdominal pain, a study of 31 of 39 abdominal computed tomographic scans in known SLE patients were suggestive of ischemic bowel (5). However, abdominal pain without evidence of active lupus tends to be caused by non-SLE-associated gastrointestinal illnesses (6). The unique aspect of this case is that abdominal pain was the only clinical manifestation of otherwise occult SLE. It was only after specific rheumatological laboratory investigations were performed during the investigation of otherwise ‘abdominal pain not yet diagnosed’ did the diagnosis of SLE become revealed. To our knowledge there have been no similar cases described in the literature. The abdominal pain was most likely secondary to either mechanical stimulation of visceral pain receptors second-
ary to the bowel edema, or due to the release of inflammatory mediators that stimulated the mucosal receptors.

Interestingly, the patient also presented with clinically mild ascites, small pleural effusions and hypoalbuminemia. Although she did have a suggestion of proteinuria on an initial urinalysis, subsequent urinalysis failed to demonstrate an ongoing renal protein loss. Therefore, we feel that in light of the bowel wall thickening and the hypoalbuminemia that there was also an SLE-associated protein-losing enteropathy (PLE).
Unfortunately, a Tc-99 labelled albumin to demonstrate protein loss from the gastrointestinal tract was unavailable, due to a lack of reagent, at the time of initial hospitalization. Of some interest, at our institution, a previous case involving a young Asian woman who was found to have a painless lupus-associated PLE as the first manifestation of otherwise occult SLE was recently reported (7). Another study reported that nine of 22 cases in the literature of autoimmune associated PLE were from Japan (8). Thus, we speculate that gastrointestinal lupus may be more common in women of Asian-Pacific descent.

Although abdominal pain is a very common complaint in patients with either an established diagnosis of SLE or classic ARA target organ damage, our experience suggests that it may be the initial clinical manifestation of otherwise occult lupus. Furthermore, we suggest that the diagnosis of lupus should be considered when a young Asian woman presents with abdominal pain and unexplained enteropathy because rapid symptomatic improvement may occur with the administration of immunosuppressant therapy.

REFERENCES
