

Ischemic necrosis with sigma perforation in a patient with Systemic Lupus Erythematosus (SLE): case report

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SUMMARY: Ischemic necrosis with sigma perforation in a patient with Systemic Lupus Erythematosus (SLE): case report.

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Systemic Lupus Erythematosus (SLE) is a chronic inflammatory rheumatic disease which affects the connective tissue. Its etiology is as yet unknown, while its pathogenesis involves the immune system.

Both genetic and environmental and hormonal factors play a key role in the impaired immune regulation. A correlation with estrogens is demonstrated by the fact that the greatest incidence is found in young women, when estrogen secretion is at its highest. The disease is also reported to worsen in women taking oral contraceptives. It is therefore believed that the components of oral contraceptives, estrogens (ethinyl estradiol) and progestins, can affect the immune profile.

Of the various complications attributed to systemic lupus erythematosus, gastrointestinal disorders are less common but potentially by far the most serious.

We report a case of ischemic necrosis with sigma perforation in a patient with SLE. Signs and symptoms of acute abdomen in patients with SLE are rare (0.2%), but serious. Most patients require an exploratory laparotomy, as the causes are often linked with vasculitis.

RIASSUNTO: Necrosi ischemica da perforazione del sigma in paziente con panvasculite da lupu eritematoso sistemico: caso clinico.

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Il Lupus Eritematoso Sistemico (LES) è una malattia reumatica infiammatoria cronica ad eziologia ancora sconosciuta e a patogenesi immunitaria. Colpisce i tessuti connettivi di tutto l'organismo.

Sono state identificate alterazioni in fattori genetici ma anche fattori ambientali ed ormonali giocano un ruolo chiave nel determinare un'alterata immunoregolazione. La correlazione con gli estrogeni viene ancor più evidenziata dal fatto che la maggiore incidenza della patologia si riscontra nelle donne giovani, quando è massima la loro secrezione. Inoltre in donne con LES è stato riportato un peggioramento della malattia durante l'assunzione dei contraccettivi orali. Queste osservazioni inducono a ritenere che i componenti dei contraccettivi orali, costituiti da estrogeni (etinilestradiolo) e composti ad attività progestinica, siano in grado di modificare l'assetto del sistema immunitario.

Tra le varie complicanze attribuite al lupus eritematoso sistemico, quelle a carico dell'apparato gastrointestinale sono meno comuni ma potenzialmente molto più gravi.

Riportiamo il caso di una necrosi ischemica del sigma con perforazione in paziente con LES. Sintomi e segni di un addome acuto sono rari in pazienti con LES (0.2%) ma gravi. Comunque la maggior parte dei pazienti con tali sintomi richiede una laparotomia esplorativa in quanto le cause sono spesso legate a fenomeni vasculitici.

KEY WORDS: Systemic lupus erythematosus - Intestinal infarction - Vasculitis - Colon perforation.
Lupus eritematoso sistemico - Infarto intestinale - Vasculite - Perforazione del colon.

Introduction

Systemic Lupus Erythematosus (SLE) is a chronic inflammatory rheumatic disease which affects the connective tissue. Its etiology is as yet unknown, while its pathogenesis involves the immune system (1). The organs most affected are the skin and the musculoskeletal, hematopoietic, digestive and cardiopulmonary systems, hence the nomenclature "systemic" (2).

LSE has been associated with changes in the genetic factors involved in modulating the immune tolerance

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of autoreactive cell clones. However, environmental and hormonal factors are also important (3). One of the most important reasons for believing that estrogens have a key role in the etiopathogenesis of SLE is that women are more vulnerable to autoimmune diseases, as they have a higher immune response than men (4). It has been calculated that the incidence of autoimmune diseases in women is nine times higher than in men. Further evidence for the correlation with estrogens comes from the fact that the greatest incidence is found in young women, when estrogen secretion is at its highest (5-11).

The use of oral contraceptives has been found to favor the onset of SLE in women without clinical or laboratory signs of the disease, and in known sufferers, a worsening of the disease while taking oral contraceptives has been reported (12-15). It is therefore believed that the components of oral contraceptives, estrogens (ethinyl estradiol) and progestins, are capable of changing the immune profile. Despite this, it should be remembered that third generation contraceptive pills contain very low doses of estrogens, the component responsible for an increased risk of thromboembolic disease (16).

Of the various complications attributed to systemic lupus erythematosus, gastrointestinal disorders are less common but potentially much more serious (17). We report a case of ischemic necrosis with sigma perforation in a patient with SLE.

Case report

A 40-year-old woman with SLE was admitted to the *Unit of Surgery and Laparoscopy Surgery* at "Incurabili" Hospital, Naples, Italy with diagnosis of abdominal obstruction. The patient, who had in the past had a left adnexectomy for ovarian cysts, had around 20 days earlier undergone a total right laparohysterectomy for uterine fibromatosis. She had suspended her SLE cortisone therapy (prednisone 25 mg/day) a few days before surgery.

On admission to our unit the patient was suffering from abdominal pain, nausea and vomiting; these signs and symptoms had already begun during her convalescence after laparohysterectomy. A physical examination on admission revealed her abdomen to be tense, while some blood tests were abnormal: CPK 309 (normal value [n.v.] <145), fibrinogen 552 (n.v. 150-450), RBC 3.71, WBC 8.9, lymphocytes 26.4 (n.v. 25-50), and proteinuria. Abdominal X-ray revealed air-fluid levels; pelvic ultrasound revealed corpuscolated film in the right abdomen and conglomerated fluid-filled intestinal loops.

Intestinal obstruction was diagnosed and the patient underwent laparotomy with adhesiolysis and debridement of the terminal ileum, as the terminal ileum was found to be anchored to the right posterior peritoneum through an omega-shaped bridle.

After 36 hours the patient presented abdominal obstruction signs and fever. Cortisone therapy was re-begun with Methylprednisolone 20 mg i.v. followed by Deltacortene Prednisone 10 mg/day i.v. Further tests were carried out: blood Albumin 3.1 (n.v. 3.5-5.2); serum nephelometry (IgG 558 [n.v. 751-1560]); C reactive protein 212 (n.v. 1-8); RA-test 52.1 (n.v. 0-25); ANA neg., AMA neg., ASMA neg., anti-LKM neg., anti-native DNA neg., ENA neg., LAC-sensitive APTT = 1.19.



Fig. 1 - Plain abdominal X-ray. Multiple air-fluid levels with distension of the intestinal loops.

A chest X-ray revealed basal consolidation and pleural effusion on both sides; plain abdominal X-ray (Fig. 1) revealed multiple air-fluid levels with distension of the intestinal loops.

Abdominal CT (Figs. 2 and 3) revealed fluid-filled intestinal loops and a gas-distended sigmoid colon with air-fluid levels.

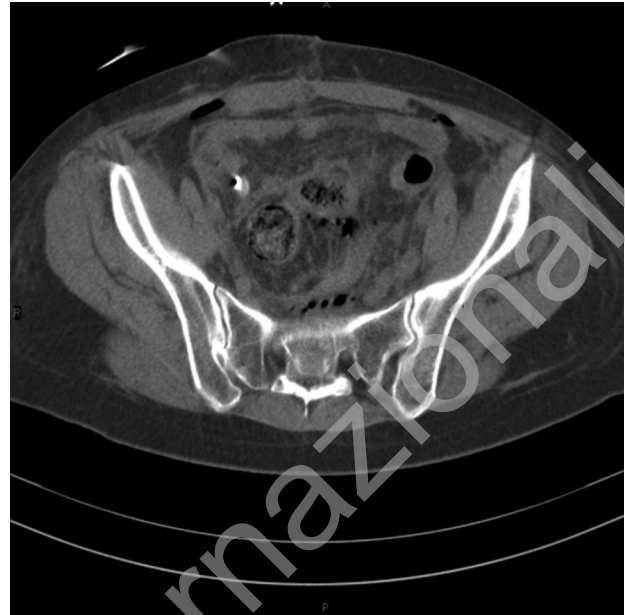
Nasogastric tube was inserted, resulting in the immediate reflux of 800 cm³ of bile fluid and gastric juices; 24 hours later the patient reported severe abdominal pain and her abdomen was even more tense. The abdominal CT was repeated (Figs. 4 and 5), this time revealing adherent, dilated fluid-filled intestinal loops. There was bilateral pleural effusion.

It was decided for re-do. Fecal material was found in the abdominal cavity, and there were recurrent weak abdominal adhesions. The mid sigmoid was perforated, while perforation due to ischemic necrosis was also incipient in the distal sigmoid (Fig. 6). The rectum was therefore mobilized down to the subperitoneal tract, followed by Hartmann's operation (TA-75 – GIA-60) with terminal iliac colostomy.

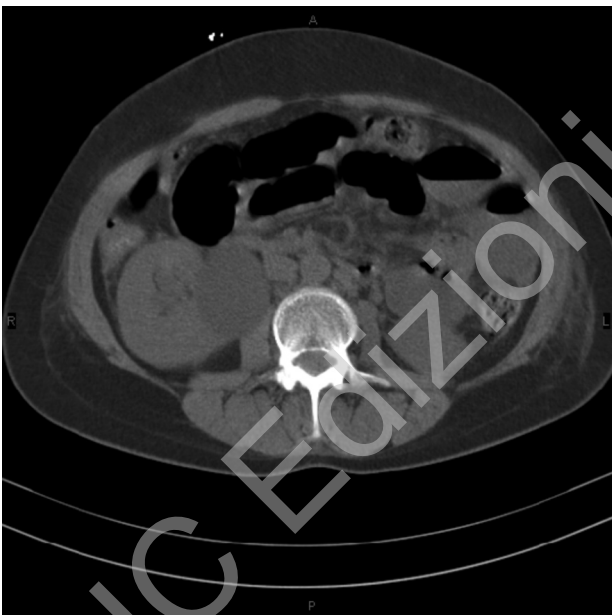
After surgery blood tests were performed: Troponin T <0.010; CK-MB 8.01 (n.v. <2.88); Myoglobin 161 (n.v. 25-58); Hb 8.5, Fibrinogen 612; gamma-GT 54; CPK up to 1415; Potassium 2.9 (n.v. 3.5-5.5); WBC 15.4; Platelets 508; D-dimers 1.8; Anticardiolipin (IgG and IgM) within normal range. The results of these and the histological examination led to the final diagnosis of ischemic necrosis with sigmoid perforation and fecal peritonitis in patient with polyarteritis nodosa secondary to SLE. After a short time in intensive care before returning to the ward, the patient was discharged 10 days after surgery with normal blood chemistry values.

Discussion

Signs and symptoms of acute abdomen in patients with SLE are rare (0.2%). Most patients require an exploratory laparotomy, as the causes are often linked to vasculitis or polyserositis, with various studies reporting a high incidence of Central Nervous System (CNS) lupus, avascular bone necrosis, thrombocytopenia and the presence of rheumatoid factor. Some patients present non-



Figs. 2 and 3 - Abdominal CT scan. Fluid-filled intestinal loops and gas-distended sigmoid colon with air-fluid levels.



Figs. 4 and 5 - Abdominal CT scan after 24 hours. Adherent, dilated and fluid-filled intestinal loops.

specific signs and symptoms of dyspepsia associated with active SLE, without any clear evidence of gastrointestinal involvement. This could in any case be linked with mesenteric vascular disease, peritoneal inflammation or certain medications. Gastrointestinal signs may take various forms: mesenteric vasculitis, esophageal disease, inflammatory intestinal disease, liver disorders or pancreatic disorders. Treatment with cortisone or immunosuppressants may also be a predisposing factor for acute abdomen.

All surgical procedures are more risky in patients with SLE than in healthy subjects, due to both the greater likelihood of complications in long-term users of corticosteroids and immunosuppressants and physical and psychological trauma related to the procedure and its possible consequences for the immune system and thus progression of the disease. When surgery is indispensable, these risks must be anticipated.

As regards our case, numerous studies have indicated that, in the presence of uterine fibromatosis, treat-



Fig. 6 - Intraoperative findings. Perforated mid sigmoid colon with incipient necrotic perforation in the distal segment.

ment with GH-SH analogs, which can induce pharmacological menopause and thus block the menorrhagia usually associated with fibroma, does not have repercussions for SLE.

Conclusions

This rare case suggests that greater attention is necessary when evaluating SLE patients with abdominal pain and abnormal blood chemical parameters who are taking or have recently suspended treatment with cortisone. The decision to perform surgery should be meticulously evaluated, bearing in mind the procedure's risks and consequences for an underlying immune disease.

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