

Patient With Systemic Lupus Erythematosus (SLE), Complex Adnexal Masses And Ascites

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Citation

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Abstract

Systemic Lupus Erythematosus (SLE) is a chronic autoimmune disease that can affect many organs. Gastrointestinal involvement is not rare and may be a cause of abdominal pain. This is often observed with ascites due to serositis involving peritoneum. Less common manifestations of lupus involving gastrointestinal tract are mesenteric vasculitis and pancreatitis. In our patient ascites is associated with laboratory findings suggesting active disease but there are also ovarian masses and elevated levels of CA125 suggesting differential diagnosis with carcinomatosis from ovarian cancer. Imaging findings (CT and US) are aspecific and need pathology report confirmation to achieve diagnosis.

INTRODUCTION

Systemic lupus erythematosus (SLE) is a rheumatologic disorder characterized by joint, skin, renal, neurologic, peritoneal/pleuropericardial and haematologic involvement. Gastrointestinal manifestations in SLE are not rare and were first described by Sir William Osler in 1895. Typical symptoms are abdominal pain, nausea, vomiting, and diarrhea. Evaluation of abdominal pain in SLE patients should consider peritoneal involvement and/or thrombotic events. In our patient abdominal pain is attributed to massive distension due to peritoneal serositis. Ascites occurs in 8–11% of patients with SLE, usually associated with nephrotic syndrome, protein-losing enteropathy, constrictive pericarditis, congestive heart failure, small vessels vasculitis or Budd-Chiari syndrome. However cirrhosis, pancreatitis, peritoneal carcinomatosis, and tuberculous peritonitis should always be considered in differential diagnosis.

CASE REPORT

A 28 year-old-woman was admitted in the Clinical Immunology Unit of the University of Pisa, with increasing abdominal distention, abdominal pain, diarrhea and dyspnoea.

A diagnosis of SLE was made ten years before, according to the 1982 ACR criteria on the basis of fever, photosensitive dermatitis, arthritis, an episode of mild pleuro-pericarditis and the presence of antinuclear and anti dsDNA antibodies (1).

Laboratory findings revealed leukopenia, hypochromic microcytic anemia, thrombocytopenia, positive direct Coombs' test, high titer of antinuclear and anti-dsDNA antibodies, low levels of C3 and C4 suggestive for SLE flare. Moreover she had also elevated levels of CA125. Abdominal US showed ascites and the presence of a mixed cystic and solid mass in the right adnexa (fig.1).

Figure 1

Figure 1: Abdominal US shows a mixed cystic and solid mass in the right adnexa.



Contrast-enhanced CT of abdomen and pelvis demonstrated

extensive septate ascites, mixed cystic and solid masses in the adnexa bilaterally 4 and 5 cm in diameter (fig.2), soft tissue thickening around the mesenteric vessels, enhancement of the parietal peritoneum and of mesenteric root (figg.3a,3b) and enlarged lymph nodes in the retroperitoneal intercavo-aortic region.

Figure 2

Figure 2: Contrast enhanced CT of the pelvis shows bilateral adnexal complex masses.

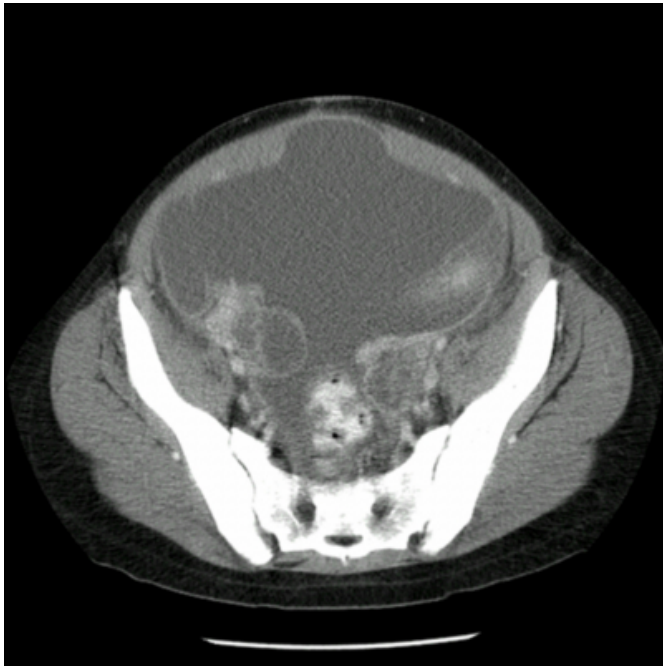


Figure 3

Figure 3ab: Contrast enhanced abdominal CT shows mesenteric thickening.

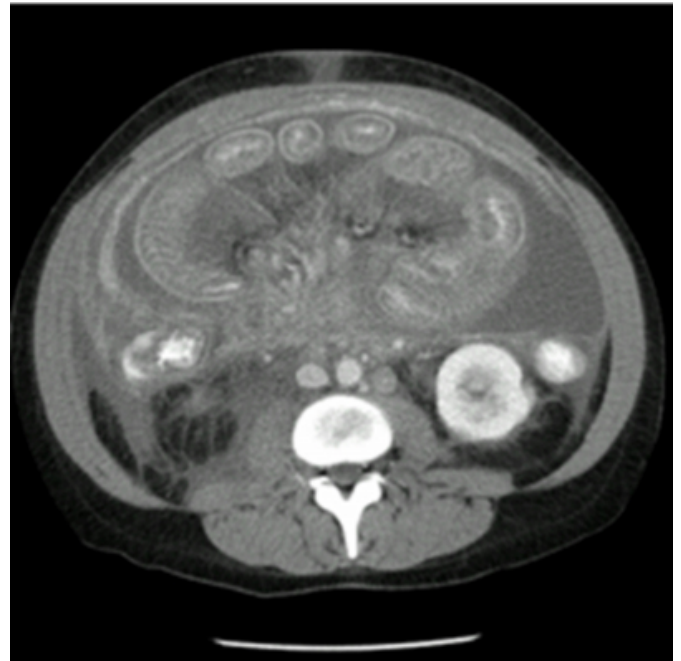
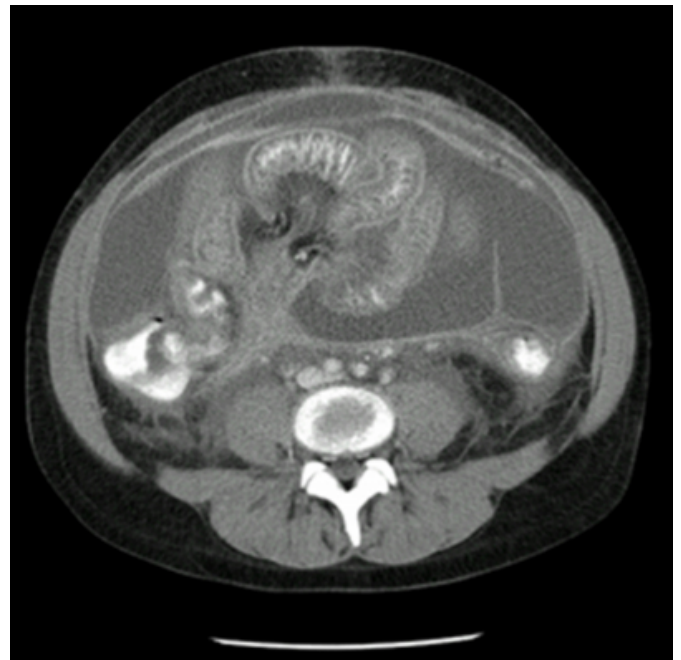


Figure 4

Figure 3ab: Contrast enhanced abdominal CT shows mesenteric thickening.



There is smooth light thickening of parietal peritoneum (3a) and of mesenteric root associated with septate ascites (3b).

She underwent also esophagogastroduodenoscopy and colonoscopy to exclude the presence of gastrointestinal

neoplasms.

Abdominal paracentesis yielded clear, yellow fluid compatible with an exudate.

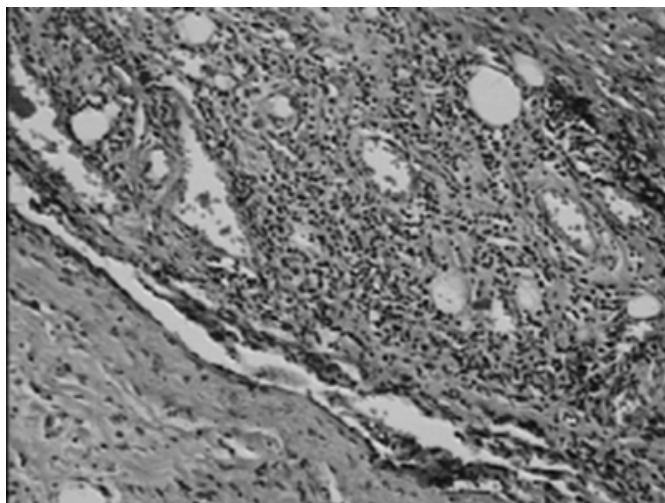
Ascitic fluid cytology and culture for bacteria and Mycobacterium Tuberculosis were negative.

Before starting with immunosuppressive therapy the patient underwent exploratory laparotomy with numerous biopsies considering in differential diagnosis ovarian neoplasms, tuberculous peritonitis and autoimmune serositis.

Peritoneal histology revealed chronic inflammation and small vessels vasculitis (fig.4). Therapy with pulse corticosteroids and cyclophosphamide resulted in dramatic disappearance of ascites which has not recurred till now.

Figure 5

Figure 4: Pathology report shows lymphocytic infiltration of peritoneum and small vessels and signs of fibrosis.



DISCUSSION

Systemic Lupus Erythematosus (SLE) is a chronic autoimmune disease of unknown origin that affects 1 in 5000 people. The disease is most common in women (female to male ratio is 9:1) of child-bearing age although it has been reported in both extremes of life. It is an autoimmune disease characterized by immune deregulation resulting in the production of antinuclear antibodies (ANA), generation of circulating immune complexes and activation of the complement system. The reasons for development of the antinuclear and other antibodies in SLE are not clear. Probably there is an association of genetic predisposition, sex hormones and environmental triggers. Disease manifestations result from recurrent vascular injury due to immune complex deposition, leukotrombosis or thrombosis.

SLE can affect many parts of the body including heart, lungs, skin, joints, blood, kidneys and nervous system. The majority of patients presents with involvement of the skin or joints usually with fever, arthralgia, Raynaud's phenomenon, photosensitive rash and alopecia. Many patients may present with internal organ involvement such as inflammatory serositis, glomerulonephritis, neurologic manifestations or hematological disorder (i.e. autoimmune haemolytic anemia or thrombocytopenia) (2,3). Gastrointestinal involvement, like in our patient, is less often observed in SLE and may be characterized by peritonitis with or without ascites due to serositis involving peritoneum (4). Another gastrointestinal manifestations of lupus is mesenteric ischemia due to vasculitis (5,6). Rare is a pancreatitis. The course of SLE is highly variable: some patients have spontaneous remissions, others respond favourably to treatment with corticosteroids and in some patients the course is unresponsive to available medications. Laboratory anomalies associated with SLE include raised erythrocyte sedimentation rate (ESR), hypergammaglobulinemia, low complement levels, anemia of chronic disease, leukopenia, thrombocytopenia, organ and non-organ specific autoantibodies. In our patient ascites was associated with a clinical history of SLE and laboratory findings suggesting active disease but she had also adnexal masses and elevated levels of CA125 that could indicate metastatic ovarian cancer too. Considering the long immunosuppressive therapy which the patient underwent, tuberculous peritonitis is also a possibility. Our abdominal CT presented some common findings in patients with SLE and acute abdominal pain, such as ascites, retroperitoneal lymphadenopathy and mesenteric edema with engorged mesenteric vessels (7) but there were also adnexal complex masses. Radiological signs were not specific, in fact ascites occurs with a variety of disease states. Peritoneal thickening is seen in SLE, ovarian cancer and tuberculous peritonitis. Usually slight, smooth peritoneal thickening with light contrast enhancement is more suggestive for benign disease, like in our case, while irregular peritoneal thickness (>2mm) with nodules and important enhancement is more suggestive for carcinomatosis (8).

US and CT showed also adnexal complex masses with thin septa but imaging appearance did not allow a conclusive diagnosis. There was also retroperitoneal lymphadenopathy that can be expression of nodal metastasis from ovarian cancer but they could be also reactive nodes due to tuberculous peritonitis or SLE. Abdominal paracentesis revealed a negative culture for Mycobacterium Tuberculosis excluding tuberculous peritonitis. Cytological examination

was negative for neoplastic cells but showed a significant increase of lymphomonocytes cells, possible expression of malignant or benign lymphoproliferative disorder such as autoimmune disease. Considering that clinical symptoms and radiological findings were not diagnostic, the patient underwent exploratory laparotomy for a definitive diagnosis. In fact before starting with pulse corticosteroid and cyclophosphamide we had to exclude the presence of malignant disease such as metastatic peritoneal carcinomatosis primarily from ovarian cancer in a patient with adnexal masses and elevated levels of CA125_(9,10). However differential diagnostic consideration have to include also peritoneal carcinomatosis from malignant neoplasms of the gastrointestinal tract and the rare primary peritoneal serous carcinoma. Pathology report revealed chronic inflammation and small vessels vasculitis of the peritoneum associated with simple ovarian cysts. The patient had a peritoneal serositis due to SLE with ascites and elevated levels of CA125. CA125 is an aspecific antigen which increases not only for malignant disease but also for inflammation involving peritoneum like in our case. Our patient had also adnexal complex masses which, associated with ascites and elevated levels of CA125, could suggest peritoneal carcinomatosis from primary ovarian neoplasm. However gynaecologic investigations including laboratory hormone findings and US imaging formed the hypothesis that they were functional cysts, then confirmed by the exploratory laparotomy.

In conclusion radiological findings in patients with SLE are aspecific and should be correlated with clinical and laboratoristic data with a close collaboration between radiologist, internist and surgeon.

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