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A Case of S L E Complicated by a Peritonitis "Lupus Peritonitis?"

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A review of the literature concerning patients with systemic lupus erythematosus reveals that the majority (about three fourths) have intestinal symptoms of some type during the course of the illness. These symptoms may be nonspecific with transient or recurrent episodes of nausea, vomiting, and diarrhea. Abdominal pain with distention, guarding, and rigidity, as seen in lupus peritonitis, can simulate many intra-abdominal syndromes. Most of these symptoms are due to vasculities of the wall of the bowel.

Lupus peritonitis is a collagen disease and frequently causes polyserositis; consequently, one might expect signs and symptoms of transient peritonitis, but some physicians never see it.

One can find only a few reports of cases which were so acute and severe as to simulate an acute abdominal surgery condition, for the cases of acute peritonitis due to lupus erythematosus are rare; but some pathologists have documented a high incidence of peritonitis at autopsy in patients who exhibited no clinical manifestation of acute peritonitis. Polyserositis is a prominent manifestation of lupus erythematosus, so the peritoneum is involved also; but serositis of the peritoneum can be at times the only manifestation of the disease.

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The following case is one in which the rare manifestations of systemic lupus erythematosus simulated an acute surgical condition of the abdomen, a condition which has been termed "lupus peritonitis".

Case Presentation

The patient was a 49-year-old white female who was brought to the emergency room by her sister because of severe and generalized abdominal pain. The history obtained at that time revealed that six days before admission she had had generalized adbominal pain and distention accompained by vomiting. Two days before admission she had consulted a private physician and was treated with "shots and pills", but she became progressively worse and was later found in a semistuporous condition.

Her past history revealed that in 1967 she had had anemia and possibly had had lupus erythematosus, but no kidney biopsy, fluorescent antinuclear antibody, or DNA complement titer were done. Lupus erythematosus test was positive and sedimentation rate was elevated. For a long period of time she had a skin rash on the hands and the abdomen; and for a while she had received low doses of steroids.

In 1968, liver biopsy had shown moderate fatty degeneration, and a diagnosis of mild diabetes mellitus was made that same year (steroid induced diabetes?). There was no previous surgery at all.

PHYSICAL EXAMINATION: Pertinent findings were: blood pressure of 70/50, pluse 110, respiration 24 per minute and shallow. Temperature was 100. Eyes: Fundi were not visualized; dry membranes. Chest: clear. Heart: midly enlarged, regular rhythm, no murmurs. Abdomen: distended, diffuse tenderness all over the abdomen. Rebound tenderness in right upper quadrant. Bowelsounds were decreased. There was flank edema. Rectal examination: negative except for occult blood, Vaginal examination was normal. Skin was the color of a red-orange peel, thickened only on the suprapubic area and both flanks. Extremities were normal with good pulsation. Neurology examination revealed only hyperactive reflexia.

LABORATORY DATA: Hemoglobin 8.7 gm%, hematocrit 26, red blood cells 2.71, white blood cells 12,400 with a shift to the right. Platelets were 104,000; prothrombin time was 12.5, control 13. SGOT 204, LDH 392, CPK 179, bilirubin 2.5 mg. Total serum protein 4.7 (albumin 2, globulin 2.7); lipase 1.4 units; cholesterol 130; amylase 91 units; glucose 225 (without i.v. running). Cl_2 was 81 mEq, CO_2 was 19.9 mEq, Na 117 mEq, K 3.9 mEq, Ca 7.3 mg., inorganic phosphours 6.8, BUN 94 mg. creatinine 3 mg. PPT 68, control 67. Blood gases showed PO_2 to be 65 mm./Hg. PCO_2 of 38 mm./Hg, PH 7.34, uric acid 12 mg. LE preparation negative; blood culture, negative, urinalysis revealed 4 + protein, 10 to 15 red cells to each magnified field. Electrocardiogram showed sinus tachycardia. Chest x-ray showed a small discoid atelectasis in the left base. Rontgenographic abdominal series showed a few loops of nondilated small bowel which contained air; findings were otherwise normal.

HOSPITAL COURSE: Twenty four hours after admission, despite i. v. fluid, Solu-Cortef, Aramine, and oxygen, the patient's blood pressure ranged between 70/50 and 100/60. I. V. Keflin and three units of blood were given. Her condition continued downhill, so an exploratory laparotomy was performed. Upon entering the abdomen, turbid fluid approximately 400 cc. was identified. Culture of this material was reported as negative. No evidence of the site of infection was found and peritoneum culture reported sterile. The spleen was markedly enlarged, and during the examination, the capsule which revealed trauma was removed. The right kidney was anomalous in shape. Needle aspiration was attempted without success. There was no evidence of organ perforation. Two days after surgery, she expired due to cardiac arrest.

DIAGNOSES BY AUTOPSY:

Adrenals:	Lipid deplection
Heart:	Dilatation; no valve disease
Lungs:	Posterobasal atelectasis
Kidneys:	Membranous glomerulonephritis consistent with lupus nephropathy
Peritoneum:	Hemoperitoneum (300 cc.), with generalized fibri-

nous peritonitis. Pleural surface smooth, no adhesions, mild thickening.

Pancreas: Pancreatic acini are dilated and contain eosinophilic secretory material. A section which included peritoneum.

Liver: Glisson's capsule is covered in some areas by a lamina of fibrous exudate. The liver parenchyma shows unremarkable histologic features.

An immunofluorescent technique was carried out on the peritoneum spicement and it was weakly positive. The spicements from spleen showed typical "onion skin lesion" of small arteries.

RESULTS: The clinical course of a particular patient with systemic lupus erythematosus manifested as lupus peritonities was presented in our study. The patient was already receiving low doses of steroide, and she had been out of medication for a long period of time. The sequence in which laboratory abnormalities preceeded clinical evidence of disease exacerbation simulated as lupus peritonitis is of particular interest.

One week prior to admission, she suffered generalized abdominal pain without any benefit of treatment. We got information of previous lupus erythematosus history from her sister when the patient was on the stretcher to go to the operating room. At the time of the patient's physical examination in the emergency room, it was revealed that this thin women had all the necessary criteria of acute peritonitis. After surgical consultation the abdominal laparatomy was carried out.

Discussion

The case history documented at this writing is the record of one of the rare cases of lupus erythematosus which involved the whole peritoneum and became so acute and severe as to simulate an acute surgical abdominal condition.

When one sees a patient with lupus erythematosus plus abdominal symptoms, one must consider these possibilities:

1. Acute lupus peritonitis

2. Lupus lesion of an intra-abdominal organ
3. Lupus with intercurrent intra-abdominal disease.

Let us review the literature with reference to each of the three conditions mentioned above.

Dubois observed gastrointestinal symptoms of lupus erythematosus in 40% of his series. Pollak et al reported on a series of 14 patients. The abdominal findings were so acute and severe as to simulate an abdomen requiring surgical intervention. The findings in 10 cases were those of vasculitis of the wall of the bowel; in four cases there was pancreatitis but no peritonitis.

Reinfenstein et al reviewed postmortem evidence of peritonitis found in 12 of 17 cases. Gormsen and Mortensen, as well as several other authors, remarked that the abdominal symptoms of systemic lupus erythematosus may be so severe as to simulate many intra-abdominal castastrophes which would warrant surgical exploration of the abdomen. The paucity of informative material in the literature reflects the rarity of this entity.

Dubois reported only one patient with acute pancreatitis; but lupus erythematosus may in time involve the serosa around the liver and spleen. For instance, Soffer and Bader noted that four of their 18 patients with lupus erythematosus experienced abdominal pain associated with perihepatitis and perisplenitis.

A study was reported by Brown et al of a series of 87 patients with gastrointestinal manifestation of lupus erythematosus who did not have acute peritonitis; but Pollak had five patients with acute lupus peritonitis in which this had been a prominent feature.

ETIOLOGY: Lupus peritomitis is frequently associated with lupus erythematous. The etiology of this manifestation remains unknown. There is no correlation epidemiologically or serologically with streptococcal infection.

The discovery of autoimmunity in lupus has provided an important clue and, as seen earlier, the auto-antibody to DNA may be incriminated as a causitive agent in glomerulitis and might be in the poluserosits. However, the biological role of autoantibodies still remains unknown.

Clinical and immunological evidence has been brought forth which suggests but does not prove the existance of a familial predisposition. It is known that intercurrent infections usually bacterial, can cause an exacerbation of quiescent lupus erythematosus or impede a therapeutic response. In lupus erythematosus, treatment of the infection alone will sometimes result in disappearance of the disease activity. Leukopenia occurs only in 57 per cent of all the patients and the rest had normal count or hyperleukocytosis, but in the case under discussion might be a part of elevation white blood cell count due to pulmonary infection.

So lupus is primarily a disease of immune complexes that circulate throughout the body and can then deposit on vascular endothelium, causing blood-vessel disease and hence can affect any organ in the body in almost any imaginable way.

PATHO-IMMUNOLOGY: Gross or microscopic evidence of peritonitis has been reported in as many as 70/75% of autopsied patients. Frequently, an underlying lupus vasculitis many be the cause of the serositis. The usual type of peritonitis seen at autopsy probably does not cause clinical symptoms, except occasionally present as in the above mentioned case report.

In this variety, there is fresh fibrinous serofibrinous or serous inflamation which can be demonstrated only microscopically. It tends to be focal, but the most common sites being perisplenic and perihepatic.

Finally, the pathogenesis of her symptoms should try to be understood. Without any doubt, the laboratory tests are most helpful in making diagnosis of flareups of lupus. Lupus is a disease in which many laboratory abnormalities may be found. An excellent and valuable test is the fluorescent antinuclear antibody or ANA. This test has been considered an extremely valuable screening test for lupus. It must be mentioned here that this test is not very specific because it is also positive in many other disease that are not related to lupus erythematosus.

A positive VDRL wihoth history of syphilis is found in about 15% of lupus cases so it is obvious that the treponema pallidum immobiliza-

tion is almost negative in these patients.

The level of complement titer will decrease in most cases of lupus These decreases have been associated with periods of activity of renal disorders or serositis. The complement level was not measured in the case under discussion.

The serologic testing can help in diagnosis of disease and also in follow-up. Changes in the serum complement levels are thought to be due to circulating antigen-antibody complexes. Recent experimental works have shown these complexes are toxic to the tissues.

The L E cell is more specific and less sensitive as a test for lupus. Probably, it occurs in about 70% or more cases of systemic lupus erythematosus. It may also be positive in other diseases.

There are some other tests which can be ordered for lupus such as rheumatoid factor, cryoglobulin, antibodies to dexyribonucleic acid (DNA) etc.

In conclusion, lupus peritonitis can simulate appendicits, cholangitis, pancreatitis, infection of the broad ligament, intestinal obstriction, paralytic ileus, and severe gastroenteritis. In the case presented herein, several differential diagnoses were made initially.

Accurate differential diagnosis is vital because surgical intervention can be the prelude to deterioration of the condition of the patient. According to those who are knowledgeable throught experience in treating lupus peritonitis, intravenous steriod therapy is extremely beneficial to the patient. So it is possible to control the exacerbation of lupus by massive doses of steriods.

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