Acute acalculous cholecystitis complicating systemic lupus erythematosus: case report and review

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Abstract

A case of acalculous cholecystitis presented as an acute abdominal emergency in a 22 year old woman with severe systemic lupus erythematosus. At the time of presentation the patient was receiving high doses of prednisone and cyclophosphamide to control her underlying disease. Histological examination of the biopsy specimen from the gall bladder showed lupus vasculitis.

This complication of systemic lupus erythematosus has not been reported before. Laboratory studies and changes in lupus activity may fail to predict the onset of cholecystitis.

Introduction

Mesenteric ischaemia, with or without bowel perforation, is reportedly the commonest acute abdominal complication of systemic lupus erythematosus, and acute pancreatitis is also well documented. Vasculitis of the gall bladder presenting as an acute abdomen occurred in a patient with giant-cell arteritis, and gall-bladder disease may also occur in polyarteritis nodosa and allergic granulomatosis.

We report a case of acute acalculous cholecystitis due to lupus vasculitis in a patient with severe systemic lupus erythematosus. This complication has not been reported before.

Case report

A 22 year old woman presented with diffuse vasculitis of the skin. Serological tests suggested systemic lupus erythematosus, and LE cells were identified. Creatinine clearance was low (normal urinary sediment), and renal biopsy showed type 4 (WHO classification) lupus nephritis. Steroids and cyclophosphamide were begun. Repeat renal biopsy six months later showed a noticeable improvement, and the steroids were subsequently reduced and cyclophosphamide stopped.

References


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She was followed up over the next two years taking 10 mg prednisone on alternate days.

In November 1981 she was admitted with severe symmetrical peripheral neuropathy and mononeuritis multiplex. Antinuclear factor was detected (1/2500), and total serum complement concentration was 106 U/ml (normal 115-150). Complement concentration remained low throughout her stay in hospital. C1q binding on admission was 100%, and three weeks later 9-3% (normal 0-5%). Antinuclear factor titre two weeks after detection was reduced to 1/500.

Because of the neurological complications prednisone was increased to 60 mg/day and methylprednisolone 3 g given intravenously. Subsequently cranial nerve complications developed, and cyclophosphamide was added. Three weeks after admission she suddenly developed severe cramping abdominal pain with associated vomiting. Examination showed tachycardia, pronounced tenderness with guarding and rebound in the epigastrium and right upper quadrant, but no fever. White cell count was 26 x 10^9/l, but serum amylase activity and abdominal radiographs were normal. The clinical diagnosis was acute steroid-induced perforation of the duodenum or cholecystitis. In view of the severe abdominal signs and white cell count laparotomy was performed.

Acute cholecystitis with severe oedema and adhesions of the gall bladder were seen. The extraperitoneal biliary system from the porta hepatitis to common bile duct was also inflamed. The gall bladder was distended but small and showed evidence of past inflammation. Because of the extent of the oedema and adhesions a cholecystostomy tube was inserted for drainage. Thick bile was aspirated from the gall bladder, and careful exploration failed to detect calculi. A full-thickness biopsy specimen of the gall bladder was taken.

Biliary contrast studies via the cholecystostomy tube 10 days later showed an irregular common bile duct with no obstruction and free flow into the duodenum. Histologically the biopsy specimen (figure) showed widespread acute inflammation which did not extend into the serosal fat. There was ulceration and necrosis of the epithelial surface, and no Aschoff-Rokitansky sinuses were seen. Mild intimal proliferation was noted in one superficial artery. In the subserosal fat the arteries showed acute inflammatory and fibrinoid change with destruction of the normal elastic medial pattern with evidence of periarterial fibrosis. Recanalisation of thrombosed vessels was evident. The vasculitis was consistent with systemic lupus erythematosus.

Removal of the cholecystostomy tube after two weeks left a persistent external biliary fistula from which about 100 ml bile drained daily. A fistulogram performed down the tract showed free communication with the biliary tree and again excluded obstruction of the common bile duct with free flow into the duodenum. Medication was reduced to 40 mg prednisone and 50 mg cyclophosphamide daily, and the fistulous tract closed spontaneously after two months. The patient had no further episodes of pain.

Discussion

Deposition of immune complexes within blood vessel walls produces acute vasculitis. Such episodes result in ischaemia and fibrosis within the target organ. In our patient at the time of the acute event the serum circulating immune complexes were only slightly raised (9-3%). Three weeks before the manifestation of acute cholecystitis, however, C1q binding was 100%. These values were therefore unreliable as an indicator of the acute event. Other indicators of lupus activity—for example, antinuclear factor and serum complement—were not helpful in predicting the onset of cholecystitis.

The pronounced irregularity of the common bile duct seen on contrast radiology may have been due to previous subclinical vasculitic episodes affecting the delicate intramural capillary network.

Few patients undergo laparotomy for acute abdominal complications of systemic lupus erythematosus. Once bowel perforation has been excluded (radiography and paracentesis) a therapeutic trial of high-dose steroids and antibiotics has been recommended. Pollack et al. used high-dose steroids successfully in five patients with severe abdominal symptoms. Our patient illustrates the limitations of such treatment, for she nevertheless required surgical intervention because of unremitting abdominal signs.

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References


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