Severe acute pancreatitis associated with peliosis hepatis in a patient with systemic lupus erythematosus

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Summary

Acute pancreatitis is an unusual complication of systemic lupus erythematosus but can also stem from immunosuppressive therapy. Although abnormal liver tests are commonly seen in SLE, peliosis hepatis is very rarely described. We report here the first case of SLE associating a severe acute pancreatitis with peliosis hepatis who responded well to the immunosuppressive therapy. As suggested by the favourable outcome in this case, the presence of peliosis hepatis in SLE cannot not be held as a strong argument against an aggressive immunosuppressive therapy. (Acta gastroenterol. belg., **2001**, 64, **298-300**).

Key words : peliosis hepatis, systemic lupus erythematosus, acute pancreatitis, treatment.

Introduction

In patients with systemic lupus erythematosus (SLE), liver function tests are often abnormal (1,2). However in most cases, liver morphology, assessed by radiological means, appears normal or shows only minor abnormalities such as an enlargement (1). Peliosis hepatis, rarely described in SLE, is characterized by blood-filled cavities in the hepatic parenchyma and has been associated with various conditions including infectious diseases, immunosuppressive drugs toxicity, hematologic or multisystem diseases (3). The precise pathogenesis is unknown (4) although cases of Bartonella infections presenting with bacillary angiomatosis peliosis have been recently reported in immunocompromised patients5. Acute pancreatitis is also a rare complication of SLE.

We report here a very rare case of severe acute pancreatitis associated with marked peliosis hepatis in a patient with SLE who responded well to the immunosuppressive therapy.

Case report

A 56-year-old man never drinking alcohol presented with severe epigastric pain, nausea, vomiting, episodic painful testis and symetrical polyarthralgia affecting small joints of the hands and the feet. Past medical history of this patient was characterized by laparoscopic cholecystectomy for symptomatic gallbladder stones in 1994 and duodenal ulcer in 1993. Previous medication consisted in omeprazole and lorazepam only.

Physical examination was characterized by fever at 38°C, poor general condition with cachexy, distended abdomen, diffusely painful on palpation. Joints of the hands and feet were swollen and tender without deformation. Laboratory tests at admission showed major inflammatory syndrome (ESR : 66 mm/h ; CRP 10 mg/dl), leucopenia and absolute lymphopenia (870 cells/mm^3). increased pancreatic enzymes (amylase 484 U/L- normal range < 82 U/L and lipase 315 U/L-normal range < 60 U/L), positive speckled antinuclear antibody (1/2500) with positive anti-SSA antibodies and a decrease in serum complement (C3 and C4). Anti-double-strain DNA, anticardiolipin IgM and IgG antibodies, lupus anticoagulant, ANCA, anti-smooth muscle and anti LKM antibodies were all negative. Serological tests for Epstein Barr virus, Cytomegalovirus, Human Immunodeficiency virus, Hepatitis B and Hepatitis C virus, syphilis and Bartonella henselae were also negative. No hyperlipidaemia.was present.

Abdominal ultrasonography showed a fluid collection in the pelvis without gall-stones and biliary dilatation tracts. ERCP was normal excluding biliary microlithiasis . Since urine examination revealed persitant microscopic hematuria, with mild proteinuria at 200 mg/day and cellular casts in the absence of infection, a renal biopsy was performed. It showed focal glomerulonephritis with fibrocellular crescent. Immunofluorescence analysis showed capillary glomerular deposits of IgM, C3, IgA and less marked deposit of IgG and C1, compatible with the diagnosis of SLE. He thus fulfilled at least four criteria (polyarthritis, lymphopenia, urinary cell casts and nuclear antibody) necessary for the diagnosis of SLE (6). He was then given methylprednisolone (64 mg per day initially) which was tapered slowly with an excellent clinical and biological response. Eight months later, the patient stopped spontaneously his treatment and after 3 more months, he had to be readmitted for abdominal pain. Serum pancreatic tests (amylase 612 U/L, lipase 439U/L) and liver function tests (Alkaline Phosphatase : 700 U/L -normal < 240 U/L ; -Gamma GT 359 U/L -normal < 42 U/L) were again raised. Abdominal CT scanner showed intraperitoneal fluid collection, perisigmoid inflammation,

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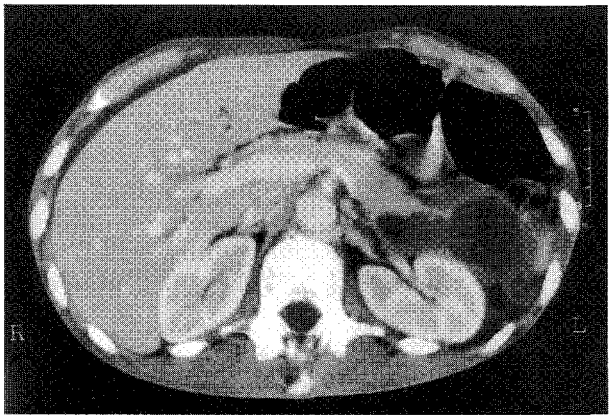

Fig. 1. — Contrast enhanced computed tomography (CT) showing pancreatic pseudocyst and normal liver morphology in a patient with SLE before treatment.

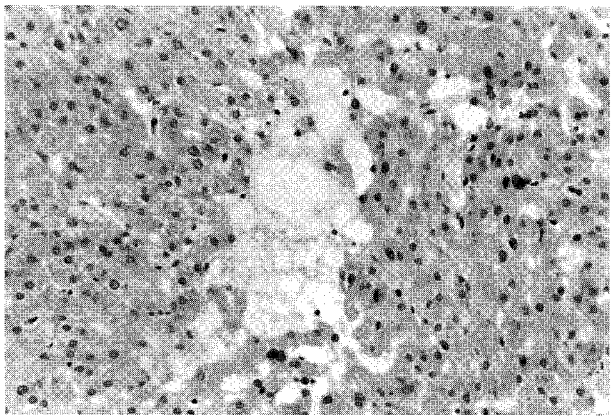

Fig. 2. — Percutaneous liver biopsy showing peliosis hepatis characterized by blood-filled cavities in the hepatic parenchyma.

thickening of mesenteric and small bowel fat and two large pancreatic pseudocysts (fig. 1). No gross morphological abnormalities could be seen in the liver. Because of unexplained bloody diarrhea, a colonoscopy was performed and revealed ulcerated and pseudopolypoid rectal lesions.

Biopsies showed non specific lesions of chronic inflammation compatible with ischemic etiology . Since there was no explanation for the abnormal liver function tests, a transcutaneous liver biopsy was performed and showed marked lesions of peliosis (fig. 2) within a normal parenchyma. The patient was treated with corticosteroids (1 g methylprednisolone during 3 days iv and then 64 mg (o.d) orally) along with azathioprine (50 mg b.d.). One week after start of treatment, a CTangiography did not show abdominal vessels abnormalities. Within 6 months, striking improvement could be observed both clinically and biologically with normalization of the liver and pancreatic function tests. One and half year later, CT scanner confirmed the disappearance of pancreatic pseudocysts. A new liver biopsy was performed by transvenous route to assess the effect of the treatment which could have potentially worsened the peliosis, still showed peliosis hepatis but at a much lesser extent than previously seen.

Discussion

We report here the first case of a patient with SLE presenting with an acute and severe pancreatitis associated with peliosis hepatis who responded well on clinical, radiological and biological grounds to immunosuppressive therapy. In SLE, symptoms related to the gastrointestinal system (e.g. nausea and vomiting) are found quite frequently (7). However acute pancreatitis is a rare complication of SLE ; it is thought to be related to inflammatory phenomenon in pancreatic arteries (8) and can be controlled with steroids (2,9). Although abnormal liver function tests are encountered commonly in SLE, peliosis hepatis is very rarely reported. To our knowledge, only two other papers have previously reported hepatic lesions of peliosis associated with SLE (1,10). However, a pathological study reveals hepatic lesions of peliosis in 6/1468 patients with SLE suggesting that peliosis hepatis in SLE is not uncommon (1). Peliosis hepatis has been associated with various conditions in particular infectious diseases, immunosuppressive states and drug toxicity, mainly with the use of azathioprine (11) or steroids (12). Cases of peliosis hepatis in renal transplant recipients have also been described (4). In patients infected with the human immunodeficiency virus, bacillary peliosis and angiomatosis have been associated with Bartonella henselae and B. quintana (5). Exact etiology of such lesions remains unknown but outflow obstruction at the junction of sinusoids and centrilobular veins with a subsequent damage to the sinusoidal barriers can explain the appeareance of peliosis hepatis (11). In our patient, pancreatic, intestinal and liver involvement suggests a disseminated vasculitis with immune complex deposits in the wall of these organs. Although corticosteroids (12) and azathioprine (11) have been involved in the development of acute pancreatitis and peliosis hepatis, the excellent outcome in our patient strongly suggests that SLE itself, through a disseminated phenomenon of vasculitis was responsible of the pancreatitis and the peliosis hepatis rather than the treatment.

Thus peliosis hepatis should perhaps be sought more often in patient with SLE with digestive involvement since these lesions, if undiagnosed and left untreated, could eventually lead to portal hypertension. Moreover, as suggested by the present case, the presence of peliosis hepatis in SLE cannot be regarded as a contra-indication for aggressive immunosuppressive therapy. Rather, if confirmed by furhter studies, an indication for it.

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