Dear Sir,

Spontaneous rupture of the liver and subsequent intraperitoneal massive hemorrhage is a rare but feared event in amyloidosis. To our knowledge, it has not been described in kidney-transplanted patients.

In 1986 a 32-year-old woman was diagnosed as having renal amyloidosis with nephrotic syndrome secondary to familial Mediterranean fever, and hemodialysis had to be started in October 1989. She received a successful cadaveric renal transplant in May 1991, and continued treatment with ciclosporine, azathioprine, prednisone and colchicine. She was also diagnosed of factor X deficiency after some episodes of mithrorrhagia. In September 1992, she was admitted because of the progressive development of fatigue, unspecific discomfort, generalized arthralgias and diathermic sensation. She had not had any recent trauma.

Physical examination revealed marked hepatosplenomegaly similar to previous explorations, without signs of ascites. There was no abdominal pain. Temperature and blood pressure were normal. Laboratory findings included serum creatinine of 1.8 mg/dl, ALT 338 U/l, GGT 591 U/l, alkaline phosphatase 300 U/l, and LDH of 999 U/l. Hemoglobin was 9.8 g/dl, leukocyte count was 1,180/µl, and platelet count was 103,000. Coagulation study showed a partial thromboplastin time of 45%; thrombin time and fibrinogen were normal. Factor X was decreased to 30% of normal value. X-ray chest film was normal.

Forty-eight hours after admission, she suddenly developed marked arterial hypotension and other signs of shock, with a rapidly increasing abdominal girth, and a severe
Spontaneous (not trauma-related) rupture of the liver is a rare event. It has been associated with primary or secondary liver malignancy, complicated pregnancy, and more infrequently, with malaria, systemic lupus erythematosus and peliosis hepatis [1]. It has been described in 6 cases associated with amyloidosis [1-5], but in a computerized search using MEDLINE we did not find a single case in kidney transplant patients. Spontaneous liver rupture in amyloidosis has occasionally been related to minimal trauma [1] or laparoscopic procedures [4]. It is usually associated with severe intra-abdominal hemorrhage, that causes fulminant death, although a case has been described decrease in plasma hemoglobin. Massive ascites was confirmed by abdominal echography. Although she was supported at the intensive Care Unit, shock was unresponsive, and 30 min later she had a cardiac arrest without response to any treatment. At autopsy, there were many pleural and pericardial adherences. Liver surface showed a rupture area (8 cm in length, 0.3 cm in width and depth) with abundant bleeding (fig. 1). In the abdominal cavity, there was a great infradiaphragmatic hematoma. Amyloid substance was found in liver, spleen, suprarenal glands and primitive kidneys. Central nervous system and kidney graft were amyloid-free.

Many factors could be involved in bleeding and rupture: (1) reduced hepatic tissue pliability secondary to amyloid infiltration; (2) increased vascular fragility as a result of infiltration of blood vessel walls [7]; (3) coagulation abnormalities, such as acquired factor X deficiency [8], increased fibrinolysis [9] or disseminated intravascular coagulation. In all cases described, death occurred within hours to days, even if the diagnosis of spontaneous liver rupture was made ante mortem [5]. In our case, liver infiltration, factor X deficiency and infiltration of blood vessel walls, all secondary to amyloidosis, could have contributed to the rupture. We can conclude that spontaneous liver rupture is an exceptional but fatal complication in amyloidosis, that can also appear in kidney transplant patients. It should be suspected in the clinical setting of sudden hypovolemia and increasing ascites. Abdominal imaging methods could produce a rapid diagnosis, but prognosis always seems to be poor.

References


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