A 28-year-old woman was admitted with severe abdominal pain and fever. She had been diagnosed as systemic lupus erythematosus (SLE) nearly 2 years previously and was under steroid treatment of 5 mg/day. She reported one medical and 2 spontaneous abortus. Recently, she was operated by cesarean section at her 40th gestational week. She developed severe abdominal pain, shaking chills and high grade fever 9 days after the operation. The liver was palpated 4 cm below the costal margin. Her right leg was tender, indurated and at the calf, its circumference was 7 cm larger then the left one. Doppler USG showed deep venous thrombosis in the right lower extremity. Laboratory studies revealed the following: WBC 23,300/mm$^3$ (%90 PNL, %10 lymphocytes), Hct 29%, platelets 67,000/mm$^3$, ALT 210 U/l, AST 212 U/l, ALP 640 U/l (N <306), total bilirubin 0.8 mg/dl, C-reactive protein 426 mg/dl (<5), ESR 135 mm/h, proteinuria 400 mg/day. CT revealed infarctions in the liver (Fig. 1) and spleen, thrombus in portal vein and mild perihepatic intraperitoneal fluid. Anticardiolipin IgG was 58.6 GPL (N: 0–8) and IgM was 60.1 MPL (N: 0–8). With the diagnosis of antiphospholipid syndrome (APS), she was started on fractionated heparin and also prednisolone of 1 mg/kg. She responded well to the therapy; prednisolone therapy was tapered, and heparin was switched to warfarin. Hydroxychloroquine was also added. A control CT obtained 1 month after the first one revealed regressed hepatic lesions, with normal spleen, and without peritoneal fluid (Fig. 2). During a follow-up of 24 months, she was doing well with normal laboratory findings.

APS is characterized by a combination of recurrent thrombosis and thrombocytopenia in the presence of elevated titers of antiphospholipid antibodies. It was first recognized in the patients with SLE and then in those with other autoimmune diseases, however it may be seen in patients without any underlying disorder. The catastrophic APS is a rare and accelerated form, represented by clinical evidence of multiple organ involvement which develops in a short time, multiple small or sometimes large vessel thrombosis, and laboratory confirmation of antiphospholipid antibodies usually in high titers. The main precipitating factors are infections, surgery, trauma, neoplasia, anticoagulation withdrawal, lupus flare, and oral contraceptives. In catastrophic APS, hepatic involvement is found in nearly one-third of the patients. In this debilitating syndrome, the optimal management is not known. The treatment of any precipitating factor, prevention or treatment of thrombotic events and suppression of excessive cytokine release are the clear aims.