LETTER

An adult patient with alcoholic liver cirrhosis and IgA vasculitis

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63-year-old man with medical history of alcohol abuse presented with abdominal pain and bloody diarrhea, weakness, asthenia, anorexia, weight loss and since two weeks a skin rash on legs and arms. Physical examination revealed a distended abdomen with positive shifting dullness, right lower quadrant abdominal tenderness, palpable purpura involving arms, legs and abdominal wall, and pitting edema of hands and feet (fig 1). Laboratory studies demonstrated normal hemoglobin and white blood cells and decreased platelet count (128.000/µL). C-reactive protein (CRP) and renal function were normal. Stigmata of active alcohol abuse and shortened prothrombin time (50%) were present. Urinalysis revealed no blood or proteinuria. CT scan abdomen showed ileocolitis and an irregular liver parenchym pattern with mosaic aspect, without porta/suprahepatic thrombosis and peritoneal fluid. Based on these findings, the tentative diagnoses of liver cirrhosis, vasculitis (possible IgA vasculitis despite negative urinary sediment) and colitis were made. Initial therapy consisted of fluid therapy, vitamin B1 and benzodiazepines. Further laboratory investigations including autoimmune and viral tests and a vasculitis work up were performed to exclude other causes of liver cirrhosis. These investigations could not reveal other causes of liver cirrhosis nor a clear explanation for the vasculitis lesions. During colonoscopy only a small polyp (dysplasia grade 3) and rectal inflammation were found. No other biopsies were taken. Gastroscopy showed esophageal varices grade II. During the next week, abdominal pain progressed and laboratory tests showed leukocytosis and increased CRP. New ultrasound revealed increased ascitic fluid and caecum and ileum wall thickening with oedema. Diagnostic paracentesis revealed a spontaneous bacterial peritonitis (SBP). Empirical treatment with amoxicillin/clavulanate was initiated. Because of clinical deterioration in the next days, antibiotics were switched to ceftriaxone and metronidazol, with resolution of the SBP. During the hospitalization vasculitis lesions were characterized by a remitting and relapsing pattern. Furosemide and spironolactone were started in combination with human albumin 20%. However, a few days later the patient developed acute renal insufficiency, and at day 30 he was transferred to our hospital. At this time new urinary analysis revealed for the first time hematuria and proteinuria of 3136 mg/24 hour. Blood results showed a slightly elevated IgA titer. Because these results made the diagnosis of IgA vasculitis more plausible, a skin biopsy was done. In the meantime treatment with angiotensin converting enzyme (ACE)-inhibition, loop diuretics and corticosteroids were initiated. Skin biopsy revealed leucocytoclastic vasculitis, with immunofluorescence microscopy demonstrating presence of

IgA immune complexes, confirming the diagnosis of IgA vasculitis. Renal function improved and proteinuria decreased. However, during the next two months the patient was recurrently readmitted with symptoms of hepatic encephalopathy, recurrent SBP and later Nocardia sepsis from a lung cavern. His clinical state further deteriorated and he died three months after first presentation.

IgA vasculitis affects predominantly children and less commonly adults. Although etiology remains unknown, IgA appears to play a key role in the pathophysiology (1-2). Clinical spectrum of the disease mainly includes cutaneous purpura, arthralgia and/or arthritis, acute enteritis and glomerulonephritis. (3). Unlike arthritis and gastrointestinal involvement, nephritis rarely precedes the onset of purpura. In fact, the onset of nephritis may be delayed for weeks or months following the appearance of other symptoms, as illustrated in our case (4). The liver has a crucial role in the clearance of intravascular IgA and cirrhosis has been associated with a variety of glomerular lesions that generally include deposits of IgA in the mesangium. Despite the frequency of glomerular IgA deposits in advanced liver disease, most adults have no clinical signs of glomerular disease (5, 6).

Table 1 shows the spectrum of liver diseases associated with IgA vasculitis reported in the literature.

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Table 1. — **Reported cases of liver cirrhosis and IgA vasculitis in the literature since 2002**

Liver disease (Author, year)	Gender, age(yrs)	Biopsy location	Clinical spectrum	Treatment	Evolution
Hepatitis C cirrhosis (Madison, 2002)	M, 63	Skin	Skin vasculitis Gastrointestinal	CS	Pneumoperitoneum Colon ischemia Died
Alcoholic cirrhosis (Miret Mas, 2003)	F, 46	Skin and kidney	Skin vasculitis Renal	NR	No complications
PBC (Gatselis, 2007)	F, 71	Skin	Skin vasculitis Gastrointestinal Renal	CS	No complications
PBC (Gatselis, 2007)	F, 72	Skin	Skin vasculitis Arthralgia	CS Methotrexate	No complications
Alcoholic/ NASH cirrhosis (Gupta; 2015)	M, 58	Skin and kidney	Skin vasculitis Gastrointestinal Renal	CS Hemodialysis	Gastrointestinal bleeding Died
Alcoholic cirrhosis (Hwang Jung, 2016)	M, 54	Kidney	Skin vasculitis Gastrointestinal Renal	CS Cyclophosphamide Hemodialysis	Gastrointestinal bleeding Died

Abbreviations: PBC, primary biliary cirrhosis ; NASH, non-alcoholic steatohepatitis ; yrs, years ; M, male ; F, female ; CS, corticosteroids. NR, not reported. *List of references can be requested to the authors.*

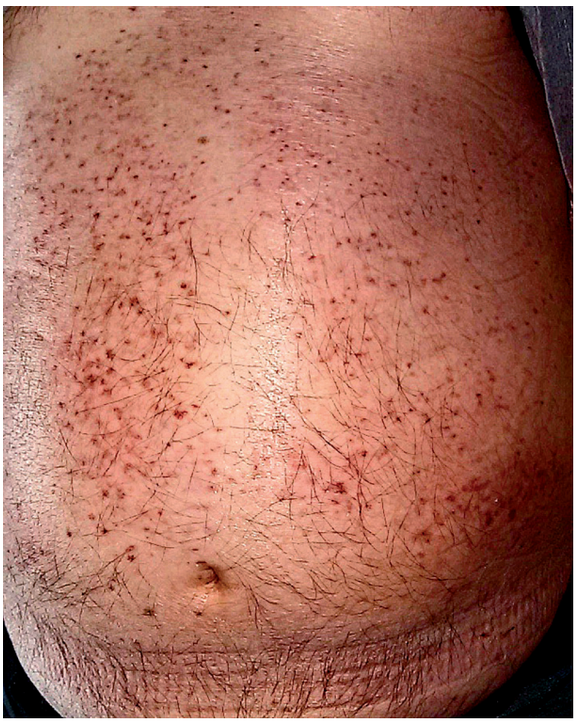

Figure 1. — Purpura and petechiae on the abdomen of the patient.

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