LETTER TO THE EDITOR

Autoimmune hemolytic anemia in a young adult with Crohn's disease and primary sclerosing cholangitis: An unusual association

KEYWORDS
Crohn's disease; Primary sclerosing cholangitis; Autoimmune hemolytic anemia

Dear Editor,

We would like to report the case of a 21-year-old patient who presented with a four-week history of bloody diarrhea with conjunctival pallor and perineal fistula orifices on physical examination. Laboratory test showed microcytic anemia (Table 1) linked to mixed origin (iron deficiency and chronic inflammation) and an anicteric cholestasis (alkaline phosphatase (PAL) 998 UI/l, gamma-glutamyl-transpeptidase (γ-GT) 160 UI/l, total bilirubin (BT) 8 mg/l). Serological markers for viral hepatitis were negative, while antinuclear antibodies and perinuclear anti-neutrophil cytoplasmic antibodies (pANCA) were positive. Endoscopy demonstrated segmental ulcerative ileo-colitis with chronic inflammation and granulomas on histological examination, consisting with CD. MRI showed segmental intrahepatic biliary ductal dilatation and multifocal strictures with normal common biliary duct. Liver biopsy revealed lymphocytic infiltration and periportal fibrosis but no biliary duct change. Given the biological and radiological features in CD context, PSC diagnosis was made. The patient was prescribed mesalazine, ursodeoxycholic acid and iron supplementation with rapid improvement (Table 1). Nine months later, bloody diarrhea relapsed associated with fever, jaundice and dark urine. There was no history of abuse of alcohol or drugs. The temperature was 38.5 °C. Laboratory tests showed normocytic anemia with hyperleucocytosis (Table 1). An anicteric cholestatic profile was noted (PAL 546 UI/l, γ-GT 96 UI/l, BT 222 mg/l, indirect bilirubin 192 mg/l). Blood cultures were sterile and there was no evidence of bile duct distension on ultrasonography. The next day, patient's hemodynamic condition worsened (blood pressure 100/50 mmHg, pulse 100 bpm) and hemoglobin level dropped (Table 1). Based on the marked reticulocytosis, indirect hyperbilirubinemia, elevated lactate dehydrogenase level (980UI/l) and decreased haptoglobin level, the diagnosis of an acute intravascular hemolysis was made. Hemoglobin electrophoresis showed normal profile. Direct Coombs Test concluded to the presence of a worm autoantibody identified as a panagglutinating IgG autoantibody. The diagnosis of an AIHA associated to a CD flare was made. Prednisone was started with good clinical and biological outcome (Table 1).

Unlike in ulcerative colitis, AIHA is an exceptional complication in CD and only 5 cases had been published, to the best of our knowledge.1–5 Interestingly, most of these cases consisted of a UC-like Crohn's colitis associated with pANCA positivity. One patient had also PSC and both diseases were diagnosed 7 years after the onset of AIHA.7 Thus, we

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Table 1 Hematologic laboratory values.

<table>
<thead>
<tr>
<th>Variables</th>
<th>First admission</th>
<th>After 3 months</th>
<th>Second admission</th>
<th>After 6 months</th>
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</thead>
<tbody>
<tr>
<td></td>
<td>Hematocrit (%)</td>
<td>Hemoglobin (g/dl)</td>
<td>Erythrocyte sedimentation rate (mm/hr)</td>
<td>Mean corpuscular volume (µm³)</td>
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<tr>
<td></td>
<td>24</td>
<td>8.4</td>
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<td></td>
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*The normal range is 16 to 199 mg/dl. **The normal range is 18 to 380 ng/ml.

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reported the second case illustrating an exceptional association of CD, PSC and AIHA but in which AIHA occurred after the onset of both CD and PSC. It’s known that AIHA can be related to an underlying disease, drug intake or can be idiopathic. Our patient had at least two underlying dysimmune diseases. However, as AIHA occurs rarely in association with these diseases, a fortuitous association of an idiopathic AIHA can’t be completely ruled out. Patient’s future evolution may further elucidate AIHA type as secondary AIHA parallels the underlying disease course.

References


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