5. Yang BL, Gu YF, Shao WJ, Chen HJ, Sun GD, Jin HY, et al. Retrorectal tumors in adults: magnetic resonance imaging findings. World J Gastroenterol. 2010;16:5822–9.
6. Gordon PH. Current status—perianal and anal canal neoplasms. Dis Colon Rectum. 1990;33:799–808.
7. Levy AD, Remotti HE, Thompson WM, Sobin LH, Miettinen M. Gastrointestinal stromal tumors: radiologic features with pathologic correlation. Radiographics. 2003;23:283–304.
8. Miettinen M, Lasota J. Succinate dehydrogenase deficient gastrointestinal stromal tumors (GISTs) – a review. Int J Biochem Cell Biol. 2014;53:514–9.
9. Joensuu H, Eriksson M, Hall KS, Hartmann JT, Pink D, Schütte J, et al. Risk factors for gastrointestinal stromal tumor recurrence in patients treated with adjuvant imatinib. Cancer. 2014;120:2325–33.

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Autoimmune hepatitis and coeliac disease. Simultaneous onset of both diseases☆

Hepatitis autoinmune y enfermedad celiaca. Aparición simultánea de las 2 enfermedades

We present a patient who developed autoimmune hepatitis (AIH) and coeliac disease simultaneously, requiring both immunosuppressant therapy and a gluten-free diet to achieve remission of both diseases.

The case concerns a 50-year-old male patient, non-smoker and occasional consumer of alcohol, with no medical or surgical prior history of interest who, after a Mediterranean cruise, developed diarrhoea without blood, mucous or pus. A complete blood count was performed that revealed hypertransaminasaemia (GPT 352 U/l, GOT 318 U/l, GGT 64 U/l), with negative serology (HAV, HCV, Epstein–Barr virus, CMV), negative stool cultures and faecal parasites, as well as normal thyroid function and iron profile. Liver enzymes were normal in a prior blood panel. The abdominal ultrasound was also normal. The patient also had dyspepsia with persistent diarrhoea. More precise lab tests were performed, which were positive for ANA (1/80) and anti-transglutaminase (248 U/ml), and revealed elevated IgG and IgA levels (18.63 g/l and 5.19 g/l, respectively). The genetic testing was positive for HLA-DR 3 and HLA-DQ 2. A gastroscopy was performed with biopsies of the second portion of the duodenum (AP: finding of intraepithelial lymphocytes and subtotal/total villous atrophy in assessed fragments of the distal duodenum) and liver biopsy (AP: periportal hepatitis with intense inflammatory activity, with moderate interface hepatitis, moderate lobular inflammatory activity with focal necrosis and fibrosis forming septa and focal architectural distortion. No steatosis or iron deposit). The pre-treatment score for AIH was 17.

After confirming the two diagnoses, treatment was started with prednisone 30 mg, azathioprine 50 mg and a gluten-free diet. The prednisone dose was gradually reduced, with normal hepatic enzyme and anti-tissue transglutaminase antibody levels four months after the introduction of treatment. IgG levels normalised at two months. After completing eight months of a tapering regimen of corticosteroids and experiencing side effects such as the onset of moon face, corticosteroid treatment was suspended, while azathioprine and the gluten-free diet were maintained. Five years after the initial onset of symptoms and following a liver biopsy that confirmed histological remission of AIH, azathioprine was finally suspended, maintaining a gluten-free diet as the only treatment. Since then, all ultrasound scans, blood tests and pathology studies have been normal. The patient is currently asymptomatic and undergoes regular check-ups for both AIH and gluten-sensitive enteropathy (coeliac disease).

It is not uncommon for a single person to have several different autoimmune diseases.[1] It has been shown that genetic predisposition can give rise to various autoimmune diseases, as they often share the same genetic loci. This is true of coeliac disease and AIH, as both conditions express similar gene combinations that encode HLA class II molecules in chromosome 6.[2] The prevalence of coeliac disease in adult patients with AIH is 10% higher than in the general population.[3] Coeliac disease has been associated with autoimmune liver diseases (AIH, primary sclerosing cholangitis, primary biliary cholangitis) as well as hepatitis B and C, non-alcoholic steatohepatitis, Wilson's disease, cirrhosis and portal hypertension and hypertransaminasaemia associated with coeliac disease.[2]

However, the unusual feature of this case is that both diseases manifested simultaneously, rather than one preceding the other (which is more common). This raises the question of whether the same trigger may have been responsible

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for two different autoimmune diseases. The simultaneous onset of both processes is less common in adults than in children.[4]

In recent years, research has been focussed on the potential benefits of a gluten-free diet to manage or improve liver enzyme levels in AIH.[5] For example, one published study that compared two patient groups found that more patients from the group with AIH and coeliac disease who started a gluten-free diet achieved disease remission (or were treatment-free) than from the group with AIH but with no coeliac disease (33% remission in the first group vs. 8% in the second).[4] Patients with AIH and coeliac disease who follow a gluten-free diet seem to be less likely to relapse after suspending immunosuppressant therapy compared to patients with AIH not associated with coeliac disease.[2,4]

As coeliac disease and AIH may occur simultaneously, the initial diagnosis of one of these processes should lead to screening for the other. Finally, following a gluten-free diet seems to have a beneficial effect on AIH in patients who have coeliac disease and AIH.

References

1. Muratori P, Fabbri A, Lalanne C, Lenzi M, Muratori l. Autoinmmune liver disease and concomitant extrahepatic autoinmune disease. Eur J Gastroenterol Hepatol. 2015;27:1175–9.
2. Narciso-Schiavon J, Schiavon LL. To screen or not to screen? Celiac antibodies in liver diseases. World J Gastroenterol. 2017;23:776–91.
3. van Gerven NM, Bakker SF, de Boer YS, Witte BI, Bontkes H, van Nieuwkerk CM, et al. Seroprevalence of celiac disease in patients with autoimmune hepatitis. Eur J Gastroenterol Hepatol. 2014;26:1104–7.
4. Nastasio S, Sciveres M, Riva S, Filippeschi IP, Vajro P, Maggiore G. Celiac disease-associated autoimmune hepatitis in childhood: long-term response to treatment. J Pediatr Gastroenterol Nutr. 2013;56:671–4.
5. Iqbal U, Chaudhary A, Karim M, Siddiqui M, Anwar H, Merrell N. Association of autoinmune hepatitis and celiac disease: role of gluten-free diet in reversing liver dysfunction. J Investig Med High Impact Case Rep. 2017;5, 232470961770567.

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Urgent liver transplantation for acute liver failure due to Wilson's disease☆

Trasplante hepático urgente por fallo hepático agudo por enfermedad de Wilson

Wilson's disease (WD) is a hereditary disorder caused by mutations of the *ATP7B* gene in chromosome 13, in which the defective biliary excretion of copper causes it to accumulate primarily in the liver and brain.[1] We present the case of a female patient with acute liver failure (ALF) due to WD.

This case concerns a 15-year-old female patient with no personal or family history of interest on treatment with oral contraceptives for some months prior, with slightly increased liver enzyme levels at baseline (ALT 89 IU/l, GGT 80 IU/l). She occasionally took non-steroidal anti-inflammatory drugs and received erythromycin on the days prior to her presenting complaint due to accidental trauma, with the development of haematoma that required drainage and the incidental finding of coagulopathy (INR 1.65), for which she was admitted to her hospital.

Clinically asymptomatic with no neurological or behavioural abnormalities (good academic record), she presented three normal haematopoietic series: INR 1.71; albumin 3 mg/dl; total bilirubin (TBIL) 1.84 mg/dl (direct bilirubin 1.22 mg/dl); AST 112 IU/l; ALT 219 IU/l; GGT 351 IU/l and polyclonal hypergammaglobulinaemia. Studies on hepatotropic viruses, autoimmunity, immunoglobulins, coeliac disease and thyroid hormones were normal. Serum copper was 36 mcg/dl (normal 80–155 mcg/dl) and ceruloplasmin 2.97 ng/dl (normal 25–63 ng/dl). The patient had no Kayser–Fleischer rings and the abdominal ultrasound revealed no pathological findings.

Progressive deterioration of liver function was observed at two weeks with a negative Coombs test for haemolytic anaemia, thrombocytopenia; INR 2.94; albumin 2.6 mg/dl; TBIL 8.35 mg/dl; ascites and splenomegaly. The patient was referred to our Liver Transplant Unit for observation and management. Upon admission, she underwent: 24-hour urine copper test of 805 mcg/dl (normal up to 60 mcg/dl) and a positive D-penicillamine challenge test.

Her liver function further deteriorated in the subsequent 48 h: INR 3.11; TBIL 25.3 mg/dl (432 μmol/l); leukocytes 10.2×10^9/l; albumin 26 g/l; AST 110 IU/l, preserved renal function, MELD score of 30 points and WD prognostic index of 13 points. In light of these findings, she was admitted to the ICU and added to the emergency liver transplant list.

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