

## Hepatic inflammatory pseudotumor related with IgG4<sup>☆</sup>



### Seudotumor inflamatorio hepático relacionado con IgG4

#### Introduction

Hepatic inflammatory pseudotumor (HIP) is a liver manifestation of a systemic disease that can occur in organs like lungs, salivary glands and pancreas. Etiology and pathogenesis of the disease is not clear with autoimmune etiology being the most common theory.<sup>1</sup> Recently, an attempt to explain the pathophysiology of the lesion in correlation with IgG4 has been undertaken. Pathological processes of inflammation can lead to specific set of changes in the tissue structure: lymphoplasmacytic inflammatory cells with fibrosis.<sup>2</sup> Herein, we report a case of HIP diagnosed by laparoscopic resection (Figs. 1–2).

#### Case report

A 60-year-old man with a 5-years history of alcohol-related chronic pancreatitis with suspected liver tumors seen on routine CT has been referred for diagnosis. CT scans showed two poorly visible hypoechoic infiltrations; one in the segment II with a diameter of 28 mm and the one in segment VI with a diameter of 20 mm. On ultrasound only the tumor in segment II with a diameter of 14 mm has been noted. The segment VI was free of any pathology on intraoperative ultrasound. Percutaneous liver fine needle biopsy has been performed with cytology showing chronic inflammatory cells. Laboratory data showed increased serum concentrations of bilirubin and white blood cells. The patient has been referred for a laparoscopic biopsy. The surgical resection of the segment II of the liver was performed by laparoscopy. On the 5th postoperative day the patient presented with abdominal pain, distention and signs of peritonitis. On emergency laparotomy bile peritonitis has been diagnosed. The leaking stump of a bile duct has been sewn and abundant peritoneal lavage performed. Further postoperative course was uneventful. Histopathological examination of the liver tumor showed diffuse inflammatory infiltration with lymphoid cells coinciding with the diagnosis of IgG4 related HIP. The blood test examination showed serum IgG4 level of 19.1 g/l.

#### Discussion

The differential diagnosis of a HIP is difficult.<sup>3</sup> On imaging studies a HIP can mimic a metastatic disease, primary liver cancer or a benign lesion.<sup>4</sup> Patients with HIP tend to be over 60 years old and more often (8×) men than women. There were several cases of exposure to industrial solvents, dust, lubricants and pesticides.<sup>1</sup> In the differential diagnosis one has to consider especially Inflammatory

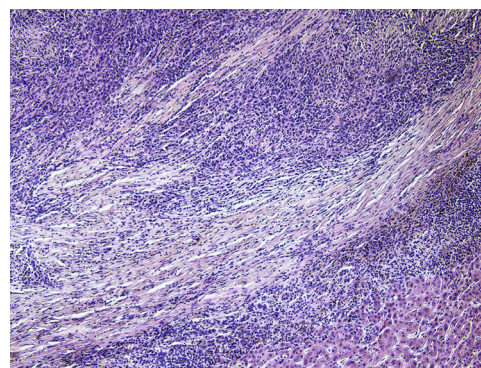


**Figure 1** Ultrasonography imaging shows a hypoechoic 14 mm mass.

pseudotumor which can occur in various locations (frequently in the lung). It can mimic malignant tumor. The lesion is composed of proliferation of fibrous tissue with presence of numerous inflammatory cells (plasma cells, lymphocytes, neutrophils, macrophages, multinucleated giant cells and eosinophils). HIP can be a IgG4-related disease. In our case there were two histological features of IgG4 – dense lymphoplasmacytic infiltrate and extensive fibrosis. On immunohistochemistry IgG stain showed many IgG-positive plasma cells and a number of IgG4-positive cells (focally >10 cells/HPF; but altogether IgG4/IgG (+) cell ratio was about 10–20%). Other tumors to be considered in differential diagnosis of HIP are tumors of parasitic origin (for example Entamoeba), neoplasms (inflammatory myofibroblastic tumor) and other causes of inflammatory infiltration in the liver.

In the majority of patients the diagnosis can be achieved by needle liver biopsy, however, as in our patient, resection is sometimes required to reach the diagnosis.<sup>5</sup>

Unfortunately, the risk of serious and even life threatening complications after this procedure is much higher than after non-surgical diagnostic procedures.<sup>6</sup> The full histopathological evaluation of the lesion, together with laboratory finding can give way to a firm diagnosis of a specific type of HIP.<sup>7</sup> Once the diagnosis is established treatment with steroids is suggested.<sup>3</sup> In patients who were diagnosed by biopsy alone, liver resection can also be required as a definitive treatment when steroid therapy is futile.<sup>5</sup>



**Figure 2** Microscopic view of IgG4-related hepatitis. The hepatic stroma shows extensive fibrosis and abundant inflammatory infiltrate.

<sup>☆</sup> This work has been presented during 6th Biennial Congress of the Asian-Pacific Hepato-Pancreato-Biliary Association in Yokohama, Japan, 7–10 June 2017.

## Conflict of interests

The authors declare no conflict of interests.

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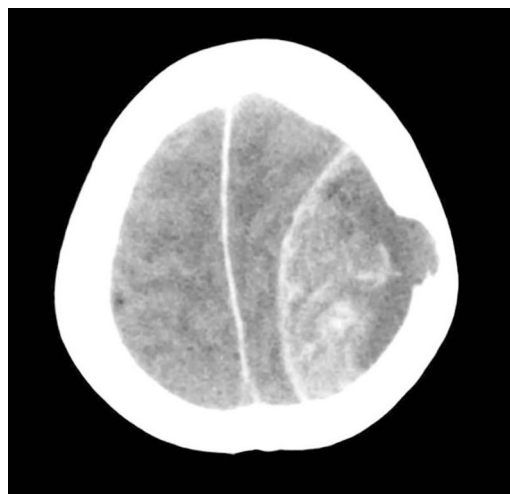
## Hematoma epidural agudo como presentación de carcinoma hepatocelular: a propósito de un caso y revisión de la literatura



### Acute epidural haematoma as a presentation of hepatocellular carcinoma: Case report and literature review

Mujer de 59 años de origen tailandés y residente en España desde hace 20 años con antecedentes personales de nefrectomía parcial e histerectomía, y que no consume fármacos de forma habitual. Acude a urgencias de nuestro hospital por cuadro de malestar general, distensión abdominal, ictericia y coluria de semanas de evolución, con empeoramiento clínico en la última semana. En la anamnesis dirigida refiere antecedente de transfusión sanguínea en los años 80. Niega consumo de fármacos, productos de herbolario ni tatuajes. Viaja frecuentemente, habiendo visitado Tailandia y Suiza en el último año. Consume 16 g/día de alcohol desde hace 5 años. A la exploración física presenta un tinte icterico cutáneo-mucoso y hepatoesplenomegalia no dolorosas. En la analítica destaca una coagulopatía no conocida previamente y alteración de los parámetros hepáticos: bilirrubina total de 8,11 mg/dl, bilirrubina directa 5,85 mg/dl, GOT 368 U/l, GPT 137 U/l, GGT 625 U/l, FA 190 U/l, PCR 14,39 mg/dl y una actividad de protrombina del 53%. Tras 24 h de ingreso presenta cuadro súbito de cefalea intensa holocraneal y deterioro brusco del nivel de consciencia (Glasgow de 5) sin focalidad neurológica. Es trasladada a cuidados intensivos, procediéndose a intubación orotraqueal y realización de

tomografía computarizada (TC) craneal con contraste intravenoso. La TC (**fig. 1**) informa de la presencia de un hematoma epidural agudo (HEA) frontoparietal izquierdo de 85 × 42 mm con fenómenos expansivos intracraneales asociados e hidrocefalia hipertensiva en ventrículo contralateral, probablemente secundario a lesión osteolítica de la bóveda craneal sugerente de etiología metastásica. La paciente es intervenida de forma urgente por neurocirugía, procediéndose a la realización de una craneotomía frontotemporal izquierda, evacuación del hematoma y resección tumoral.



**Figura 1** TC craneal: hematoma epidural agudo frontotemporal izquierdo. Lesión osteolítica en bóveda craneal de probable origen metastásico.