If the tumour is asymptomatic but the metastases cannot be resected, then it is preferable to administer chemotherapy from the outset, with a view to attempting to render the metastases resectable. However, when the tumour is symptomatic (obstruction or perforation), the recommended approach is surgery on the primary tumour followed by chemotherapy; the liver metastases can be resected in the same procedure if they are resectable.[4] In this case, given that the tumour was symptomatic, it was decided to perform surgery in a single procedure, since both metastases required minimal hepatic resections, in order to be able to prevent a second surgical procedure which might have increased the patient's risk of post-operative morbidity and mortality.

In 2016, a case report was published that reported the finding of an adenocarcinoma co-occurring with a neuroendocrine tumour in the colon; however, we were unable to find another case with two simultaneous colon lesions (one adenocarcinoma and one neuroendocrine tumour) as well as lymph node involvement and liver metastases by both tumours.[5] Thus, we present one of the first cases in the literature with two co-occurring colorectal neoplasms of different histological origins (adenocarcinoma and neuroendocrine tumour) with co-occurring respective liver metastases. This case moreover features a report of the resection of two colon neoplasms and two liver metastases in the same procedure.

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Francisco J. Tejero-Pintor*, José C. Sarmentero-Prieto, Martín Bailón-Cuadrado, José I. Blanco-Álvarez, Javier Sánchez-González, Rosalía Velasco-López, Paloma Rodriguez-Vielba, David Pacheco-Sánchez

Servicio de Cirugía General y Digestiva, Hospital Universitario Río Hortega, Valladolid, Spain

*Corresponding author.
E-mail address: fjtejeropintor@gmail.com
(F.J. Tejero-Pintor).

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Usefulness of digital cholangioscopy for IgG4-related cholangitis diagnosis and cholangiocarcinoma exclusion☆

Utilidad de la colangioscopia digital en el diagnóstico de la colangiopatía por IgG4 y en la exclusión de colangiocarcinoma

IgG4-related cholangiopathy is the most common extrapancreatic manifestation of type 1 autoimmune pancreatitis (AIP) and both fall under the umbrella of systemic IgG4-related disease.[1] It is characterised by the appearance of biliary stenosis and the most common presentation is obstructive jaundice (70–80%).[1] It is diagnosed through clinical suspicion, imaging tests, serum IgG4 levels and histological analysis.[2–4] It is important to rule out a neoplastic origin for the biliary stenosis; hence, cholangioscopy may be very useful, although there are few published data in this regard.[5] Treatment with corticosteroids may help to confirm the diagnosis if disease remission is demonstrated.[2]

We present the case of a 67-year-old patient with the following signs and symptoms for the past two months: steatorrhoeic faeces, early satiety and weight loss. A computed tomography scan showed ''sausage pancreas'' with peripheral ring enhancement. This typical image, together with serum IgG4 levels twice the upper limit of normal (3110 mg/l [80–1400 mg/l]), confirmed a diagnosis of type 1 AIP[3,4] with associated exocrine pancreatic insufficiency. Treatment was therefore started with pancreatic enzymes.

After remaining asymptomatic for 10 months, he went in for epigastric pain which had started three days previously, choluria and 5 kg of weight loss. Laboratory testing revealed increased cholestatic enzymes and transaminases. Three weeks later, his signs and symptoms as well as his laboratory values had improved, and his IgG4 levels were normal (631 mg/l). He underwent magnetic resonance cholangiopancreatography (MRCP) which showed a filiform main pancreatic duct and dilation of the intrahepatic bile duct with no representation of the common hepatic duct (CHD) and adjacent enhancement (Fig. 1). Cholangiocarcinoma could not be ruled out.

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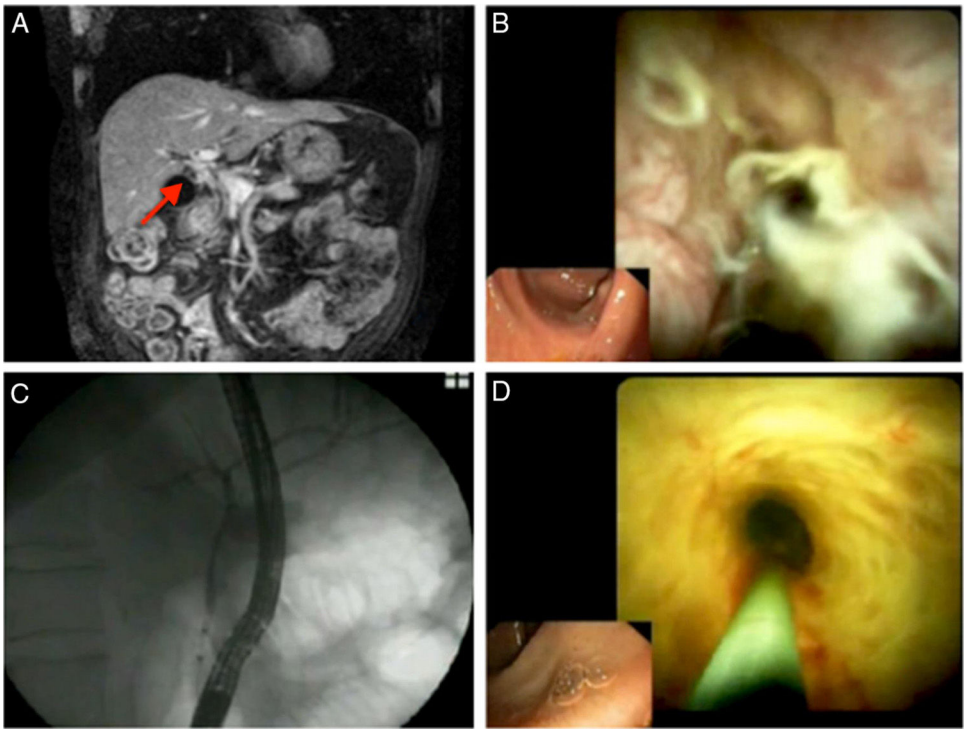

Fig. 1 (A) Magnetic resonance cholangiopancreatography showing the absence of representation of the common bile duct with contrast enhancement adjacent to its path. (B) First cholangioscopy: image of the common bile duct showing an epithelium with an inflammatory appearance, with diffuse erythema, a papillary–granular pattern and dilated, tortuous vessels. (C) Normal cholangiogram following treatment with corticosteroids. (D) Cholangioscopic image of the common bile duct following treatment showing a ''honeycomb'' pattern with depressed areas having a fibrotic appearance resulting from scarring.

Endoscopic retrograde cholangiopancreatography (ERCP) revealed several stenoses with dilation of the intrahepatic ducts and filiform stenosis in the CHD. Cytology detected epithelial cells with atypical nuclei initially suggestive of cholangiocarcinoma, although they may also be seen in other inflammatory processes. Ultimately, following review by two pathologists, a diagnosis of cholangiocarcinoma could not be confirmed.

Given the suspicion of cholangiopathy associated with AIP, it was decided to start treatment with oral prednisone (0.6 mg/kg/day). This achieved complete resolution of symptoms and normal laboratory values. However, to rule out cholangiocarcinoma entirely, a cholangioscopy (SpyGlass DS®, Boston Scientific) was performed when treatment was started. This showed the CHD and common bile duct to have erythematous walls, with a papillary–granular pattern and dilated, tortuous vessels (Fig. 1), with a soft consistency on biopsy (SpyBite®, Boston Scientific). Pathology analysis confirmed the absence of malignancy and demonstrated the presence of severe lymphoplasmacytic infiltration, fibrosis of the lamina propria and scant plasma cells with IgG4 (two per high-power field [HPF]).

Four months later, after the cycle of corticosteroids had been completed, ERCP (SpyGlass DS®) was repeated and revealed a strictly normal cholangiogram with disappearance of stenosis and a common bile duct with a ''honeycomb'' image, with a fibrotic–scarring appearance (Fig. 1).

The presence of biliary stenosis, the prior diagnosis of type 1 AIP and the patient's improvement with corticosteroids confirmed a diagnosis of IgG4-related cholangiopathy.[2]

IgG4-related cholangiopathy should be included in the differential diagnosis in cases of masses and biliary stenosis, in which histological analysis is essential to rule out cholangiocarcinoma.[2] Initially, most samples were obtained from surgical procedures due to suspicion of tumour disease; now there is greater disease identification and it is possible to perform a cholangioscopy, which enables bile duct biopsies to be taken.[5]

However, histological confirmation, which requires more than 10 IgG4-positive cells per HPF, may be difficult. There are still limitations due to discontinuous biliary involvement, insufficient samples and a drop in IgG4-positive cells in stages of fibrosis and following treatment.[1]

To arrive at a diagnosis of IgG4-related cholangiopathy, it is important to correlate histological findings with the patient's signs and symptoms, serology and imaging tests. If the other criteria are fulfilled, then histological confirmation is not essential for diagnosis. However, it is necessary to rule out cholangiocarcinoma; hence, cholangioscopy plays an important role.

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Ana Sanahuja Martínez*, Isabel Pascual Moreno, Andrés Peña Aldea, Vicente Sánchiz Soler, Francisco Mora Miguel

Servicio de Medicina Digestiva, Hospital Clínico Universitario de Valencia, Universitat de València, Valencia, Spain

*Corresponding author.
E-mail address: anasanahujamar@gmail.com (A. Sanahuja Martínez).
2444-3824/

First case report of pancreatic myeloid sarcoma diagnosed through EUS-FNA

Primer caso clínico de sarcoma mieloide pancreático diagnosticado mediante AAF-EE

Endoscopic ultrasound fine-needle aspiration (EUS-FNA) plays a major role in gastrointestinal neoplasia staging. It also proved to be an effective technique in the differential diagnosis of pancreatic lesions allowing tissue sampling for pathology evaluation.[1]

The authors report a 74 year-old male, admitted due to epigastric pain, weight loss and vomiting. His past medical history included ischaemic heart disease, hypertension, diabetes mellitus, chronic renal failure and prostatic cancer. No alcohol consumption or previous episodes of acute pancreatitis were present. The patient underwent an abdominal magnetic resonance which revealed a large, ill-defined, contrast-enhanced pancreatic neck-body mass with diffusion restriction (Fig. 1A). Laboratory evaluation was unremarkable except for mild anaemia. Serum amylase and CA19.9 was normal. Gastroenterology consultation was required and the patient was scheduled for an EUS. A 20 mm heterogeneous eccentric pancreatic body mass extending to the retroperitoneum was observed. The lesion displayed positive Doppler sign (fine flow) with celiac trunk abutment although without evidence for vascular invasion. FNA through the stomach using a 25 gauge needle (two passes) was performed with air-suction aspiration (10 cc and 5 cc) (Fig. 1B and C). No lymph nodes were noted. Cytopathology identified cells with large granular cytoplasm and peripheral nuclei with prominent round nucleolus. Immunohistochemistry was positive for CD34, CD68, vimentin and CD71, being negative for myeloperoxidase (MPO) and for the usual epithelial and neuroendocrine markers (Fig. 1D–F). The diagnosis of a primary pancreatic MS was assumed. Acute myeloid leukaemia/bone marrow infiltration was excluded. After haematology consultation the patient was proposed for chemotherapy but died few weeks after the diagnosis due to rapid disease progression and decompensation of his medical comorbidities.

Myeloid sarcoma (MS), also known as granulocytic sarcoma or chloroma, is a solid haematological neoplasia composed by immature myeloid progenitors located outside the bone marrow. This rare condition may occur simultaneously with acute myeloid leukaemia as an extramedullary disease manifestation in 2–10% of the cases, although it may precede its diagnosis or be observed when the disease relapse after treatment. Isolated MS is atypical, being characterized by absence of bone barrow and peripheral blood involvement. It may affect the digestive system leading to common misdiagnosis.[2,3]

The first case of pancreatic MS was described in 1987. Actually, these pancreatic lesions are extremely rare with approximately 17 cases reported in literature and only 7 presenting as an isolated condition. Clinical manifestations are usually unspecific and radiological features often mimic pancreatitis or pancreatic adenocarcinoma. Normal tumour markers and routine blood tests are usual. MS diagnosis remains challenging and pathological analysis with immunohistochemistry is crucial to differentiate MS from other pancreatic neoplasms, often requiring an expert pathologist. Positive immunostaining for MPO, CD 68, CD 45, CD 34 and/or CD 43 is common. Although MPO has a high sensitivity and specificity for myeloid cells, being an important marker for MS diagnosis, some authors reported its expression only in 66% of the cases, which is also dependent on the type of sampling and the differentiation of neoplastic cells.[2–5]

Chemotherapy and radiotherapy are valid therapies however, the prognosis is generally poor.[4]

In previous described cases of pancreatic MS, diagnosis was suspected by radiologic imaging and in some patients only performed after surgical resection. To the best of our knowledge, a definite diagnosis through EUS-FNA had never been accomplished. This case report describes the EUS features of this uncommon disease and proves the accuracy of the cytopathological diagnosis in this clinical setting. The routine preoperative characterization of solid pancreatic lesions through EUS-FNA must be considered aiming to avoid useless surgeries and futile therapeutic interventions.