mass); biopsy-proven peripheral lymphadenopathy (submandibular, iliac or axillary) in 3 (6%); extensive small bowel mesenteric involvement in 1; and pulmonary involvement in 1 patient. Exocrine insufficiency occurred in 23 patients (44%) and diabetes in 17 (33%). Portal and/or splenic vein thrombosis developed in 7 patients (14%). 45 patients (87%) received steroids for a median of 4.6 months (range 0.6–76). A good response to steroids was seen in 43 (96%) of these patients. Disease relapse occurred in 24 (46%), with a median time to relapse of 4.2 months (range 0–38). Extrapancreatic disease at presentation predicted relapse ($P = 0.0004$), but elevated serum IgG4 did not ($P = 0.14$). 19 patients (37%) received additional immunosuppression (azathioprine ($n = 17$), mycophenylate ($n = 1$), ciclosporin ($n = 1$), methotrexate ($n = 1$), infliximab ($n = 1$)). All patients who relapsed responded to subsequent treatment. 37 patients required biliary stenting with a mean of 2 stents (range 1–10 stents). Three patients (6%) died during follow-up, all from probable IgG4-related disease: autoimmune encephalitis, progressive liver disease and hilar cholangiocarcinoma.

Conclusion Long-term follow-up in this large cohort shows that AIP/IgG4 systemic disease may be associated with multi-organ complications, which cause significant morbidity, and a small but significant mortality. Extrapancreatic involvement at presentation predicts a complicated course.

Competing interests None.

OC-015

DISEASE PROFILE AND LONG-TERM OUTCOME OF PATIENTS WITH AUTOIMMUNE PANCREATITIS/IGG4 SYSTEMIC DISEASE

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Introduction The diagnostic features, disease course, and management of autoimmune pancreatitis (AIP)/IgG4 systemic disease are still being defined. Here we report long-term data on the largest European cohort of prospectively followed patients.

Methods All patients referred to our centre from 2004–2010, in whom a diagnosis of AIP was made, were followed prospectively.

Results 52 patients (41 M, 11 F; median age 59 years (range 26–85)) were included, with a median follow-up from diagnosis of 32 months (range 0–76). 40 patients were jaundiced at presentation (77%) and 7 patients (14%) had undergone surgery for suspected pancreaticobiliary cancer. HISORT criteria were fulfilled in 46 (89%) patients. Serum IgG4 was raised in 35 patients (67%). Histological samples were taken in 49 patients. Tissue IgG4 was raised (>10 cells/hpf) in 29 patients (59% of those who had at least one biopsy). 17 patients had associated autoimmune disease (7 Sjögren's, 7 thyroid disease, 4 IBD and 1 vitiligo). 41 patients (79%) had extrapancreatic disease: biliary involvement in 37 patients (71%) (of whom 2 (4%) developed cirrhosis and 1 (2%) underwent liver transplantation); retroperitoneal fibrosis in 5 patients (10%); renal infiltrates in 5 (10%); arthropathy/arthritis in 3 (6%); IgG4-related cerebral involvement in 2 patients (4%) (encephalitis and pituitary