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A unique clinical phenotype of primary sclerosing cholangitis associated with Crohn's disease ☆

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Abstract

Background and aims: A distinct clinical phenotype has been demonstrated for ulcerative colitis with concomitant primary sclerosing cholangitis (PSC). The course and behaviour of Crohn's disease (CD) with PSC has, in contrast, never been defined. We aimed to define the characteristics of patients with concomitant PSC and CD.
Methods: The Oxford PSC and IBD databases were abstracted for: PSC subtype, date of diagnosis, symptom onset, smoking history, Mayo Clinic PSC score and outcomes (hepatic failure, liver transplantation, Montréal CD classification, treatment, cancer and death). Patients with PSC/CD were matched 1:2 to two control groups: one with PSC/UC and one with isolated CD.
Results: 240 patients with PSC were identified; 32 (13%) with CD, 129 (54%) with co-existing UC, and 79 had PSC without IBD. For PSC/CD vs. CD controls, isolated ileal CD was less common (6% vs. 31%, $p=0.03$). Smoking was less common in PSC/CD (13% vs. 34%, $p=0.045$). No difference in the distribution of CD, or treatment required was observed. For PSC/CD vs. PSC/UC controls, more patients with PSC/CD were female (50% vs. 28%, $p=0.021$). 22% of PSC/CD patients had small duct PSC compared with 6% with PSC/UC, ($p=0.038$). Major event-free survival was prolonged in the PSC/CD group compared with PSC/UC, (Cox regression $p=0.04$).

Abbreviations: CD, Crohn's disease; IBD, Inflammatory bowel disease; OLT, Liver transplantation; PSC, Primary sclerosing cholangitis; UC, Ulcerative colitis; UDCA, Ursodeoxycholic acid; 5-ASA, 5-aminosalicylic acid

☆ Author contributions: JD, ML and EC assisted JSH with data collection and review. JSH drafted the manuscript and designed the study together with RWC. SPLT and RC critically appraised the manuscript.

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