Primary Sclerosing Cholangitis Developing Years After Ulcerative Colitis: A Rare but Likely Underrecognized Phenomenon

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Introduction:
Primary sclerosing cholangitis (PSC) is a chronic liver disease characterized by inflammation and fibrosis of intrahepatic and extrahepatic bile ducts, which can progress to cirrhosis and liver failure. More than 70% of patients with PSC have concomitant inflammatory bowel disease (IBD), most commonly ulcerative colitis (UC). In contrast, the prevalence of PSC among patients with UC is much lower, estimated at 2.5%. This association may be underrecognized, however, as subclinical PSC is increasingly identified in patients with long-standing UC.

Case:
A 68-year-old woman with a history of ulcerative pancolitis diagnosed in 2005, complicated by poorly differentiated sigmoid adenocarcinoma requiring proctocolectomy with ileal pouch-anal anastomosis (IPAA), was evaluated for persistently elevated liver enzymes 18 years after her UC diagnosis. A magnetic resonance imaging and a magnetic resonance cholangiopancreatography (MRI/MRCP) revealed diffuse intrahepatic and extrahepatic biliary dilation extending to the peripheral ducts, consistent with PSC. Notably, an MRI/MRCP performed one year ago showed only mild intrahepatic ductal dilation without extrahepatic involvement. She remains clinically stable on mesalamine suppositories three times weekly and ursodiol 600 mg twice daily, with ongoing follow-up in hepatology and IBD clinics. Surveillance MRI/MRCP is planned every six months to monitor disease progression or complications.

Discussion:
PSC is an uncommon but clinically significant comorbidity in patients with UC and may remain undetected for years due to subclinical disease. The literature suggests that PSC can develop long before clinical, biochemical, or radiographic abnormalities become apparent, raising the potential for delayed or missed diagnoses. Our case illustrates this phenomenon, with PSC identified nearly two decades after the patient’s UC diagnosis. This prompts the question of whether PSC is truly rare in long-standing UC or simply underrecognized. Importantly, this case highlights the need for ongoing surveillance for PSC in patients with UC, even when symptoms and laboratory abnormalities are absent, as early recognition may help prevent serious complications such as cirrhosis and malignancy.