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doi:10.1016/j.gie.2008.03.1095

Sclerosing cholangitis from microscopic polyarteritis: an 8-year follow-up case report

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Disruption to the arterial blood supply of the biliary epithelium can lead to ischemic cholangitis, with ensuing strictures, cholestasis, and, if unresolved, progression to secondary sclerosing cholangitis. Reported causes include iatrogenic hepatic arterial injury, hepatic artery thrombosis in liver transplantation recipients, or intra-arterial administration of chemotherapeutic drugs.¹ Rarely, ischemic cholangitis can develop as a result of vasculitis affecting the mesenteric blood vessels. We present a case that shows the progression of ischemic cholangitis in a patient with microscopic polyarteritis (MPA) who developed symptomatic sequential common bile duct (CBD) and hilar strictures. Side-by-side plastic stents and bilateral hilar stents were used for the treatment of CBD and hilar strictures, respectively, in conjunction with cytotoxic and immunosuppressive therapy.

CASE REPORT

A 37-year-old woman with no medical history presented with epigastric and right upper-quadrant pain. She underwent a series of investigations that included upper and lower endoscopies, US, abdominal CT, hepatobiliary iminodiacetic acid scan, and laparoscopy, all of which were unremarkable. Subsequently, a further severe episode of abdominal pain was associated with mild mixed abnormalities of liver function tests (LFT) and biliary dilatation. An

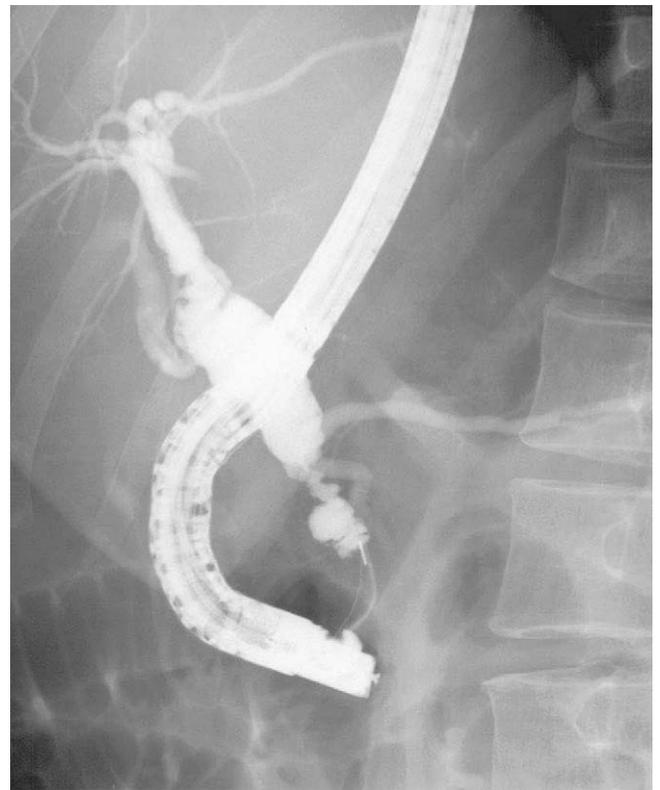


Figure 1. Finding on ERCP, showing a distal CBD stricture with proximal biliary dilatation.



Figure 2. Finding on ERCP, showing resolution of distal CBD stricture but diffuse abnormality with irregular ridging of the biliary wall and pseudodiverticuli.

ERCP was performed. The papilla and pancreatic duct were normal. The bile duct had a pleated configuration at the distal end, with a stricture in the mid CBD and proximal dilatation. The intrahepatic ducts were normal (Fig. 1). A 10F, 7-cm-long plastic stent was inserted. A subsequent exhaustive investigation, including an EUS, failed to discern an etiology. A subsequent ERCP at 3 months showed improvement of the biliary stricture, and she began a program of side-by-side stenting. The stricture and the proximal dilatation resolved, although the cholangiogram revealed an irregular ridged biliary wall and pseudodiverticuli (Fig. 2). The patient was now asymptomatic and declined further follow-up.

Four years after the initial ERCP, she developed polyarthrititis, peripheral neuropathy, and cutaneous vasculitis, with symptoms of cholestasis and abnormal LFTs. An ERCP revealed bilateral hilar strictures with dilatation, which was more pronounced on the right. There was also retraction of the biliary tree, with shortening of the CBD (Fig. 3A and B). Antinuclear antibodies were positive (1 in 320). Perinuclear antineutrophil cytoplasm antibodies (p-ANCA) were also positive. Erythrocyte sedimentation rate was elevated at 96 mm/h (normal = 0-26 mm/h). Abdominal angiogram performed on 2 separate occasions revealed microaneurysm formation. The changes predominantly involved the renal and hepatic arteries, although

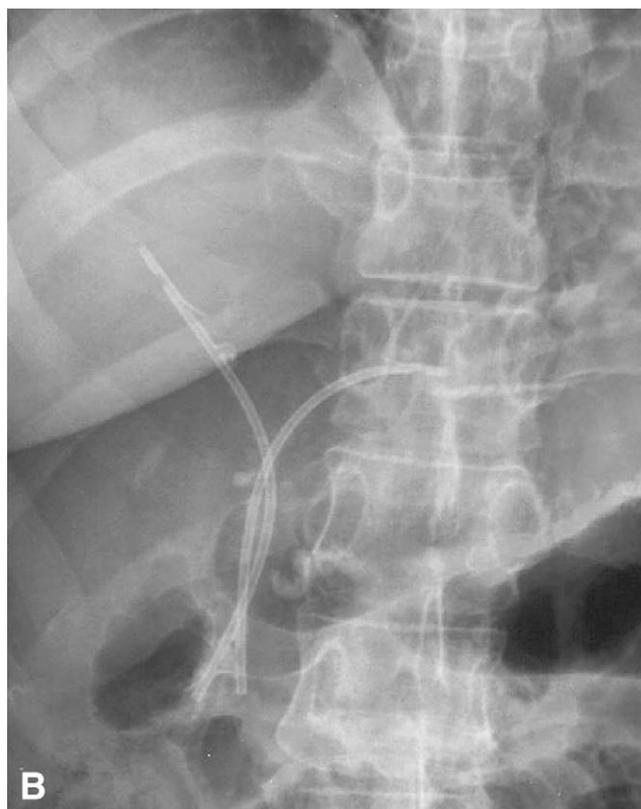
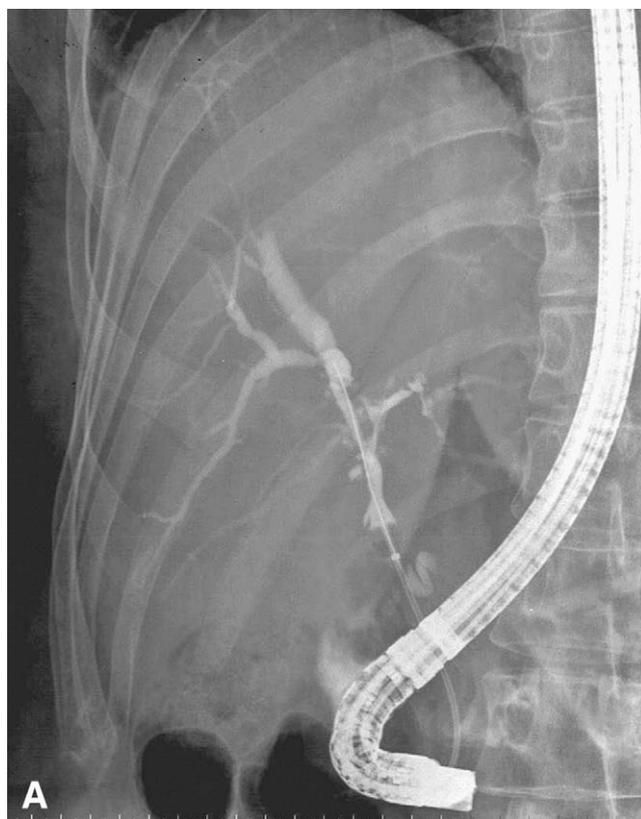


Figure 3. A, Balloon occlusion cholangiogram, showing shortening of the bile duct, bilateral hilar strictures, and proximal dilatation. **B,** Bilateral 7F hilar stents were placed.

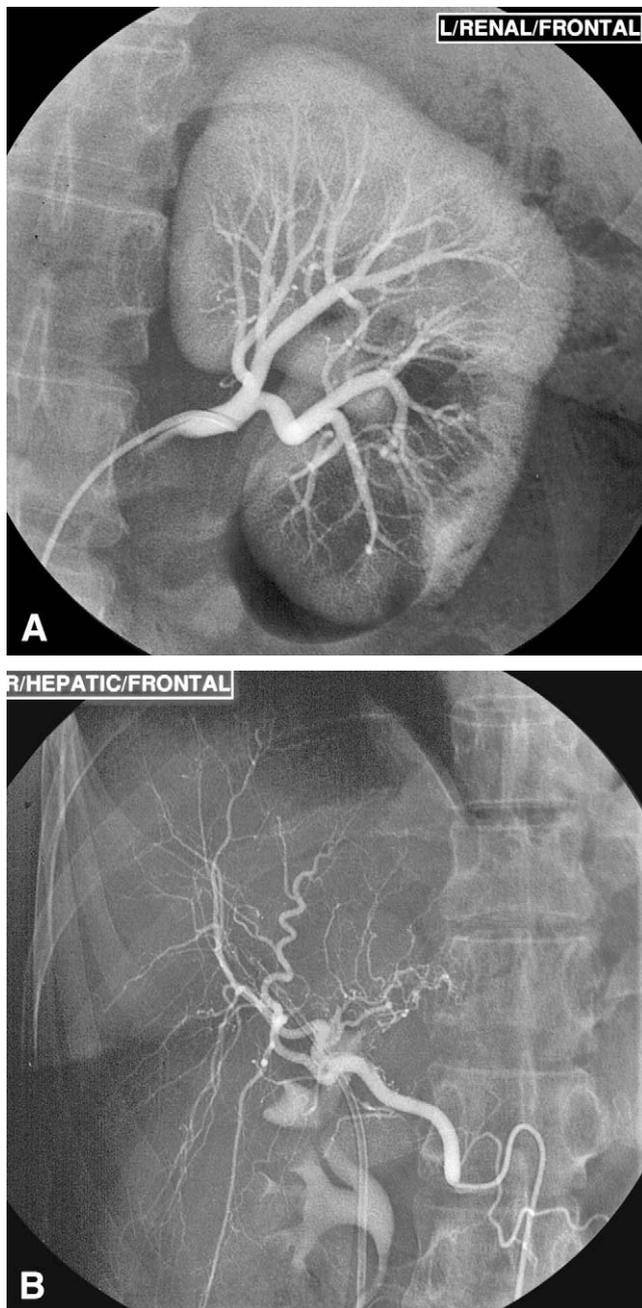


Figure 4. Findings on abdominal angiogram. **A**, Microaneurysm formation affecting the branches of renal arteries. **B**, Microaneurysm formation affecting the branches of hepatic arteries.

renal function remained normal. (Fig. 4A and B). The diagnosis of MPA was made. The patient was treated with pulse methylprednisolone and cyclophosphamide, followed by tapering of oral prednisolone, which was later replaced by oral methotrexate.

Bilateral 7F hilar stents were used for 12 months. The gross cholangiographic abnormalities persisted; however, the hilar and CBD strictures resolved (Fig. 5). Biochemical evidence of cholestasis and symptoms resolved, and she has remained well for the past 3 years.



Figure 5. Finding on ERCP 8 years after presentation; biliary strictures are absent.

DISCUSSION

Biliary involvement has been described in patients with polyarteritis nodosa (PAN), Wegener's granulomatosis, leukocytoclastic or hypersensitivity vasculitis, and Schönlein-Henoch purpura.¹⁻⁴ To our knowledge, it has not been reported in patients with MPA, which is a necrotizing small-vessel vasculitis that often affects venules, capillaries, arterioles, and small arteries. It is distinct from classic PAN, which does not usually involve small vessels. In addition, patients with MPA are often serologically positive for p-ANCA, unlike those with PAN.

The vascular supply of the bile duct is derived from a rich peribiliary network of vessels that stem from the hepatic artery. In our patient, the biliary changes were most likely because of progressive involvement of the vascular supply of the bile duct, which led to ischemia and inflammation, with subsequent fibrosis. In patients with vasculitis and biliary involvement, histologic assessment revealed arterial occlusion from intimal fibrinoid necrosis, thrombosis, and granulation tissue.² Selective damage to the bile duct has also been seen in situations of compromised arterial hepatic flow, such as in patients who are critically ill, and, if untreated, can progress to secondary sclerosing cholangitis.⁵

In conclusion, this case demonstrates the evolution of biliary injury from MPA and its subsequent improvement with cytotoxic and anti-inflammatory therapies in conjunction with biliary and hilar stent placements. Biliary involvement in systemic vasculitis, although rare, should be

included in the differential diagnosis of patients who are seen with unusual cholangiographic findings and cholestasis.

DISCLOSURE

All authors disclosed no financial relationships relevant to this publication.

Abbreviations: CBD, common bile duct; LFT, liver function test; MPA, microscopic polyarteritis; p-ANCA, perinuclear antineutrophil cytoplasm antibodies; PAN, polyarteritis nodosa.

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doi:10.1016/j.gie.2008.03.1125

Gastric-duplication cyst with an aberrant pancreatic-ductal system: an unusual cause of recurrent abdominal pain

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Gastric duplication cysts (GDCs) are very rare in adults. Reported here is a case of a GDC communicating with an accessory pancreas via an aberrant duct in a 49-year-old woman with recurrent abdominal pain.

CASE REPORT

A 49-year-old woman was seen at an outside hospital with epigastric pain. An EGD revealed an antral mass. On a CT of the abdomen, a cystic lesion of the antrum was seen. She was referred to our institution. Another EGD revealed an antral submucosal mass with a central umbilication (Fig. 1). EUS revealed a 37-mm × 26-mm submucosal cyst suggestive of GDC (Fig. 2). However, results from the aspiration of the cyst revealed a high amylase level. Her symptoms subsided but recurred 4 months later. EUS confirmed cyst reaccumulation, and aspiration again showed a high amylase level. No malignant cells were seen. After aspiration, her symptoms improved again. Because of her recurrent symptoms, an endoscopic retrograde pancreatography (ERP) was performed, which revealed a second pancreatic-duct system that originated from the main pancreatic duct near the tail of the pancreas (Fig. 3). This second system extended parallel to the main pancreatic duct before attaching to the antral cyst. The patient went for surgery, and the accessory pancreas, aberrant pancreatic duct,



Figure 1. An upper endoscopy image, showing an antral submucosal mass with a central umbilication.

and cystic portion of the antrum were resected. The surgical specimen showed a 5-cm round structure identified as a cyst, which was attached to a 17-cm-long accessory pancreas with the aberrant ductal system (Fig. 4). The cyst was unilocular, with smooth lining. On microscopic examination, the cyst wall was lined by gastric mucosa with focal