Think Outside the Duct, to ID Mirizzi Syndrome

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LOS ANGELES — Mirizzi syndrome is an uncommon cause of bile duct obstruction, but one that it is vital to recognize, David L. Carr-Locke, M.D., said at the 12th International Symposium on Pancreatic and Biliary Endoscopy sponsored by the Cedars-Sinai Medical Center.

That's because the condition requires management quite different from that used for the two conditions it mimics: choledocholithiasis and neoplastic obstruction, said Dr. Carr-Locke, director of endoscopy at Brigham and Women's Hospital, Boston.

As opposed to bile duct obstruction caused by an internal stone, Mirizzi syndrome is defined as duct obstruction due to external compression from a stone or inflammation in the infundibulum of the gallbladder or a cholecystocholedochal fistula, said Dr. Carr-Locke, also of Harvard Medical School, Boston.

Symptoms of the syndrome mimic simple bile duct stones and strictures; patients may have abdominal pain, obstructive jaundice, and cholangitis.

Symptoms of the syndrome—named after Argentinian surgeon Pablo Luis Mirizzi—bear a frustrating similarity to symptoms of simple bile duct stones and strictures. Patients may present with abdominal pain, obstructive jaundice, and cholangitis.

"You can't see a patient in your office and say, 'ah, yes, this is Mirizzi syndrome,' " Dr. Carr-Locke said.

Imaging remains the key to detecting Mirizzi syndrome. One clue is anatomic. A short cystic duct, a low origin of the cystic duct, or a cystic duct that runs parallel to the common hepatic duct may predispose to the syndrome, he said.

On the CT scan, an inflammatory mass around the stone may resemble cancer. Magnetic resonance cholangiopancreatography may actually demonstrate extrinsic compression.

More often, though, suspicions arise during endoscopic retrograde cholangiopancreatography, with the appearance of normal pancreatic ducts and what looks like a "rather square, piston-like stone," at the neck of the gallbladder or "in" the bile duct, Dr. Carr-Locke said.

"When you try to remove it, it will not budge," he said. "Contrast will not flow all the way around the stone."

"I would emphasize repeatedly, if the stone doesn't move, there has to be a reason for it," he added.

In its simplest form, Mirizzi involves extrinsic compression caused by a stone or inflammation originating in the infundibulum of the gallbladder, or, more rarely, in a long cystic duct remnant after cholecystectomy or a mucocele of the cystic duct after liver transplantation. This is known as type I Mirizzi syndrome.

Various classification systems attempt to categorize more complex forms of the syndrome, specifically, stones entwined in cholecystocholedochal fistulas arising from necrosis that can develop along the adjacent walls between the bile duct and gallbladder. Such stones can be lodged partly within the bile duct and partly within the cystic duct or gallbladder.

One system Dr. Carr-Locke considers helpful comes from Chile and categorizes type II Mirizzi syndrome as a cholecystobiliary fistula that involves less than a third of the bile duct circumference at the time of surgery. Type III Mirizzi syndrome involves two-thirds of the bile duct, and type IV indicates complete bile duct destruction (Br. J. Surg. 1989;76:1139-43).

"For many of these patients, the treatment is surgical," Dr. Carr-Locke said.

It can be quite helpful if an endoscopist can describe the degree of bile duct destruction along the lines of the Chilean classification system because an expert biliary surgeon may need to be recruited for reconstruction of a badly damaged duct.

In any case, the interventional endoscopist's role in treating Mirizzi syndrome is to relieve the obstruction by stenting or draining to stabilize the patient.

Occasionally, it may be possible to fragment the offending stone using direct laser or electrohydraulic lithotripsy in patients awaiting surgery or those who are poor surgery candidates, he noted. ■