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Primary Biliary Cholangitis vs Primary Sclerosing Cholangitis: Do You Know the Difference?

Announcer:
Welcome to CME on ReachMD. This episode is part of our MinuteCE curriculum.

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Dr. Mayo:
This is CME on ReachMD, and I'm Dr. Marlyn Mayo. Here with me today is Dr. Kris Kowdley.

Dr. Kowdley, can you explain the differences between primary biliary cholangitis [PBC] and primary sclerosing cholangitis [PSC] in terms of pathology and diagnosis? And in particular, what should providers be considering in their differential diagnosis?

Dr. Kowdley:
Yes, so thank you. The diagnosis of primary biliary cholangitis and primary sclerosing cholangitis can seem confusing because there are some similarities between the 2 conditions, but they are very, very different. So, just to begin, primary biliary cholangitis is clearly an autoimmune disease that predominantly affects women – 90% of patients are women, although 10% or thereabouts can be men affected by PBC. PSC, by contrast, has a somewhat male predominance.

The most important difference between these 2 conditions is that PBC is a condition that affects the small, microscopic bile ducts within the liver that are referred to as interlobular bile ducts. They're generally 80 microns in size or smaller. And so the damage to the bile ducts is really something that's only seen on liver biopsy or reflected in blood liver biochemical tests. The major bile ducts, in other words, the large bile ducts – the common bile duct, the common hepatic duct, etc. – which you can see on a cholangiogram, obtained either by MRI or endoscopy, in PBC, are generally normal.

By contrast, PSC affects these large bile ducts in the liver, and so the diagnosis requires, in most cases, a cholangiogram showing a characteristic abnormality of the bile ducts which is often referred to as a beaded appearance with saccular dilatations and strictures, and these can be seen throughout the bile ducts that are visible to the naked eye.

Another really key important difference between PSC and PBC is the association with inflammatory bowel disease. So primary biliary cholangitis, usually not associated with IBD, or inflammatory bowel disease, but PSC, very closely linked to inflammatory bowel disease, and this may be either Crohn's colitis or ulcerative colitis.

By contrast, when you look at patients who have primary sclerosing cholangitis, or PSC, about 50% to 80% of patients, if examined carefully with a colonoscopy and biopsies, will actually have evidence, either visibly or on biopsy in the colon, of inflammatory bowel disease.

So in summary, there is a similarity in how the patients present in that they both present with a cholestatic pattern of liver abnormalities, which for us means that the alkaline phosphatase, gamma GT are elevated out of proportion to the aminotransferases, meaning the AST and the ALT. The complications of the liver disease are similar in both cases. In cases where the disease has progressed to an advanced stage, patients may develop cirrhosis and complications of liver disease.

Dr. Mayo:
Thanks. So it sounds like there are some differences between PBC and PSC in terms of the associated conditions, but the major difference between the 2 is the size of the ducts affected, and that translates into a key difference in how you make the diagnosis, with PSC typically affecting the larger ducts and requiring a cholangiogram and PBC affecting the smaller ducts and not being visible on cholangiogram.

Thank you very much, Dr. Kowdley. Thank you to our listeners. I hope that you learned something from this discussion today.

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