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Released: 11/08/2024
Valid until: 11/08/2025
Time needed to complete: 54m

ReachMD
www.reachmd.com
info@reachmd.com
(866) 423-7849

Does Your Patient Have PBC?

Announcer:
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Dr. Kowdley:
Hi, this is CME on ReachMD, and I'm Dr. Kris Kowdley. Today I'll discuss best practices when assessing patients with suspected primary biliary cholangitis, or PBC.

Here are the topics we'll cover: initial clinical assessment of a patient with suspected PBC, the symptoms that the patients may present with, associated common hepatic and extrahepatic manifestations, and how do we confirm a diagnosis of PBC.

So patients with PBC may often present with asymptomatic elevation of liver test abnormalities or may present with symptoms, which I'll talk a little bit more about subsequently. And the symptoms may be fatigue or pruritus most commonly.

The liver test abnormalities may be detected during routine screening for health assessment or in evaluation of symptoms. The characteristic liver test abnormalities in PBC are what we call a cholestatic pattern, which is an elevation of alkaline phosphatase and gamma-GT out of proportion to the amino transferases, which is ALT and AST. And usually the ratio is 2:1 for alkaline phosphatase to AST and ALT.

Now, it is important, when you see a patient who has this type of a presentation, to take a very good history for past medical history and concomitant medications, because medications can sometimes lead to elevated liver tests from a very similar pattern.

Now, the symptoms of PBC may include fatigue and pruritus. And often the fatigue and pruritus are not recognized until we specifically ask the patient about those symptoms. So it's very important to quantify the degree of pruritus and try and assess whether it's mild, moderate, or severe; exacerbating factors, whether there's any association with climate, weather, or other contact stimulation.

With regard to the extrahepatic manifestations of PBC, there could be quite a lot. PBC is an autoimmune disease, and there are other autoimmune diseases that are commonly associated with PBC. These include thyroid disease, which is seen in about 30% to 40% of patients; bone disease, particularly osteopenia and osteoporosis in postmenopausal women can be a very common symptom; and patients may frequently have what we call sicca syndrome, dry eyes and dry mouth. They may have this in the absence of clear-cut Sjögren's syndrome, and up to 40% of patients may have dry eyes and dry mouth.

So it's important to take a comprehensive history in assessing the patient with suspected PBC in the context of elevated alkaline phosphatase and gamma-GT out of proportion to AST/ALT. It's important to get some sort of imaging exam to make sure the bile ducts are not dilated to rule out a concomitant biliary tract process.

Now, the hepatic complications of PBC are really no different from any other chronic liver disease, and patients with advanced disease may present with variceal hemorrhage, spontaneous bacterial peritonitis, jaundice, muscle wasting, etc. But our goal, of course, is to

identify patients well before the development of these complications.

Now, how do we confirm the diagnosis of PBC? This is a really important point, because a liver biopsy is not needed for the diagnosis of PBC. If 2 out of 3 criteria are met, namely a cholestatic pattern of liver test abnormalities, a positive antimitochondrial antibody and sufficiently high titer, or a disease-specific ANA, which is the sp100 and gp210 antibody for ANA – if any of these 3 are present, then you can confirm the diagnosis of PBC, and a liver biopsy is not needed. A liver biopsy is only needed for the remaining 3% to 4% of patients who do not have these autoantibodies.

So in summary, PBC should be suspected in a patient with chronic cholestatic liver disease, and can be confirmed with the presence of a positive AMA or disease-specific ANA.

Thank you.

Announcer:_
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