Cholangiocarcinoma Clinical Presentation

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History

Signs and symptoms of cholangiocarcinoma include the following:

- Jaundice
- Clay-colored stools
- Bilirubinuria (dark urine)
- Pruritus
- Weight loss
- Abdominal pain

Jaundice is the most common manifestation of bile duct cancer and, in general, is best detected in direct sunlight. The obstruction and subsequent cholestasis tend to occur early if the tumor is located in the common bile duct or common hepatic duct. Jaundice often occurs later in perihilar or intrahepatic tumors and is often a marker of advanced disease. The excess of conjugated bilirubin is associated with bilirubinuria and acholic stools.

Pruritus usually is preceded by jaundice, but itching may be the initial symptom of cholangiocarcinoma. Pruritus may be related to circulating bile acids.

Weight loss is a variable finding. It may be present in one third of patients at the time of diagnosis.

Abdominal pain is relatively common in advanced disease. It often is described as a dull ache in the right upper quadrant.

Physical

If the cholangiocarcinoma is located distal to the cystic duct takeoff, the patient may have a palpable gallbladder, which is commonly known as Courvoisier sign.

An abdominal mass or palpable lymphadenopathy is uncommon, but hepatomegaly may be noted in as many as 25% of patients.

Causes

The etiology of most bile duct cancers remains undetermined. Currently, gallstones are not believed to increase the risk of cholangiocarcinoma. Chronic viral hepatitis and cirrhosis also do not appear to be risk factors.

Infections

In Southeast Asia, chronic infections with liver flukes (Clonorchis sinensis and Opisthorchis viverrini) have been causally related to cholangiocarcinoma.

Other parasites, such as Ascaris lumbricoides, have been implicated in the pathogenesis of cholangiocarcinoma.

Observations have raised the possibility that bacterial infections with Helicobacter species may play an etiologic role in biliary cancer.[9]

Inflammatory bowel disease

A strong relationship exists between cholangiocarcinoma and primary sclerosing cholangitis. Cholangiocarcinoma generally develops in patients with long-standing ulcerative colitis and primary sclerosing cholangitis.[10] The lifetime risk of developing this cancer in the setting of primary sclerosing cholangitis is 10-20%. At increased risk are patients with ulcerative colitis without symptomatic primary sclerosing cholangitis and a small subset of patients with Crohn disease.

Chemical exposures

Certain chemical exposures have been implicated in the development of bile duct cancers, primarily in workers in the aircraft, rubber, and wood-finishing industries.

Cholangiocarcinoma has developed decades after administration of the radiologic contrast medium thorium dioxide (ie, Thorotrast). This product, which results in lifelong alpha particle irradiation by thorium decay products, was in use from the 1930s until the 1950s.[11]

Miscellaneous conditions

Congenital diseases of the biliary tree, including choledochal cysts and Caroli disease, have been associated with cholangiocarcinoma.

Other conditions rarely associated with cholangiocarcinoma include bile duct adenomas, biliary papillomatosis, and alpha₁-antitrypsin deficiency. Obesity may also be a risk factor.[12]

Differential Diagnoses

http://emedicine.medscape.com/article/277393-clinical
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