Biliary Rhabdomyosarcoma
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History
2 year old male with jaundice.

Diagnosis
Biliary Rhabdomyosarcoma

Discussion
Embryonal rhabdomyosarcoma of the biliary tree, sometimes referred to as boytroid rhabdomyosarcoma, only accounts for 0.04% of childhood neoplasms (1). It accounts for 1% of all embryonal rhabdomyosarcomas (1). The median age of presentation is 3 years with a slight male preponderance (2). The tumor often exceeds 8 centimeters at the time of discovery and can invade the duodenum (2, 3). It can arise from almost anywhere along the biliary tree including liver, intrahepatic and extrahepatic biliary ducts, gallbladder, or ampulla (4). It has also been reported to arise from hepatic and choledochal cysts (4).

The most common clinical features are jaundice and abdominal distention with pain, vomiting, and fever being less frequent (2, 5). Elevation in liver transaminases and bilirubin is often present. A tumor arising from the biliary tree discovered in children over one year of age is most commonly an embryonal rhabdomyosarcoma, however other considerations include: choledochal cysts, inflammatory pseudotumor, and cholangiocarcinoma arising within a choledocal cyst (4). In older children, considerations would also include hepatoblastoma and hepatocellular carcinoma, which would be distinguished from embryonal rhabdomyosarcoma by elevated alpha fetoprotein levels (1). Ultrasonography generally demonstrates biliary ductal dilatation, an intraductal mass, or fluid filled mass if the tumor has a cystic component (1). Because of this, it can have a radiologic appearance similar to a choledochal cyst, especially if there is no local invasion (3). CT may show a heterogeneous or hypointensuating mass with biliary ductal dilatation (4). MRI has advantages over other modalities because of its ability to define the extent of disease and relationship to hepatic vasculature (1). Generally, intraoperative or percutaneous tissue sampling is needed for definitive diagnosis (7). Surveillance monitoring should be performed for many years after resection due to the possibility of late recurrence (4). CT may be the best modality for surveillance of recurrence (1).
A review of the literature has reported several different surgical approaches depending on the extent of the tumor burden. These include pylorus sparing pancreaticoduodenectomy (1, 8), Roux-en-Y hepaticojejunostomy (9), and choledochojejunostomy with cholecystectomy and end to side jejunojunostomy (6) to name a few. In some cases, chemotherapy and radiation therapy may be the initial treatment to shrink tumor burden prior to surgical consideration (1). Because of the use of adjuvant chemotherapy, positive margins after surgical resection are tolerable and do not seem to increase mortality. With the combination of therapies, survival has increased significantly and a recent study reported a survival rate of greater than 75%, compared to 25% in 1970 (6). However, other studies recommend longer term follow up studies because hepatobiliary rhabdomyosarcoma has been reported to recur up to 9 years after therapy (4).

Findings
US-Solid and cystic mass in the porta hepatis with biliary dilation.
CT-Solid and cystic mass in the porta hepatis with biliary dilation.
MR-Solid and cystic mass in the porta hepatis with biliary dilation.

Reference

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