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Splenic Hemangiomas

Vascular tumors of the spleen in infants and children include in order of higher frequency hemangiomas, littoral cell angiomas, angiosarcomas, and hamartomas. Splenic hemangiomas are rare benign tumors that present clinically with enlargement of the spleen, thrombocytopenia and microangiopathic hemolytic anemia. Initial work-up should include ultrasound and computed tomography scans of the abdomen. MRI is very helpful since can diagnose the hemangioma and help it differentiate from a hamartoma of the spleen. At MRI splenic hemangiomas showed signal intensity characteristics and enhancement patterns similar to those described for other body hemangiomas, while hamartomas demonstrated diffuse heterogeneous enhancement on images obtained early after administration of contrast material and became more uniformly enhanced on delayed images. Scintigraphic studies are mandatory to confirm diagnosis. Primary hemangiosarcoma of the spleen is an aggressive neoplasia which occurs in a previously splenic benign hemangioma. In such cases children present with symptoms of recurrent fever, malaise, and abdominal pain. Management consists of total splenectomy. Should the hemangioma be massive preoperative splenic artery embolization can be achieved. Antiangiogenic therapy has been reported efficacious in large lesions.

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Pancreatic Carcinoma

Tumors of the pancreas can arise from exocrine or endocrine cells. Adenocarcinoma of the pancreas is an extremely rare tumor found during the pediatric age. Most adenocarcinomas of the pancreas are non-islet cell lesions of ductal origin with more than 70% located in the head of the pancreas. Others are acinar cell carcinomas and nonfunctional islet cell

carcinomas. More than half the cases are females with a mean age of nine years (range three months to 18 years). As in adults this is a very aggressive malignant neoplasm with early metastatic spread to lymph nodes and liver. While the majority of pancreatic tumors are exocrine lesions of ductal origin, acinar cell tumors are more commonly observed in children and associated with a better prognosis. Children with pancreatic ductal carcinoma presents with abdominal and back pain, vomiting, obstructive jaundice, a palpable mass and weight loss. Diagnostic imaging should consist of ultrasound, CT-Scan and MRI to look for chances of surgical resectability. Genetic markers such as CEA, C19-9, and alpha fetoprotein should be obtained. Whenever feasible and in the event of no metastatic disease management should consist of surgical resection of the tumor. Body and tail tumors can be dealt with distal pancreatectomy, while head of pancreas tumors will need pancreaticoduodenectomy (Whipple procedure). The response rate with chemotherapy and/or radiotherapy is very poor. The prognosis with metastatic disease is dismal.

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Hepatic Lymphangiomas

Lymphangiomas found within the liver are an extremely rare finding exclusively present in children and adolescent. Most reported cases occur as part of a diffuse body involvement with lymphangioma including spleen, kidneys, gastrointestinal tract, mediastinum and lung. Histologically the lesion presents with multiple smooth-walled cysts filled with serous fluid and blood looking like multiple intercommunicating cavernous spaces separated by thin-walled septa and lined by a flattened endothelium. Children develop hepatomegaly and right upper quadrant pain. Lymphangiomas are benign lesion, but within the liver they can produce symptoms due to progressive compression of vital structures. Diagnostic imaging consists of ultrasound, CT-Scan and MRI. Management consists of resection if a surgical liver anatomic lesion or pedunculated is identified, while total hepatectomy and orthotopic liver transplantation is used with symptomatic diffuse hepatic involvement. Sclerotherapy utilized in other lymphangiomas in the rest of the body is of no benefit with hepatic lesions.

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