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Imaging features of pancreatoblastoma

Abstract  Background. Pancreatoblastoma is a rare tumour of childhood. Reports of the imaging appearances are limited.  
Objective. To define the imaging features of pancreatoblastoma by analysis of four previously unreported cases and review of the literature.  
Materials and methods. Findings at CT (n = 4), US (n = 3) and MRI (n = 2) were retrospectively reviewed in four patients with pancreatoblastoma. A Medline search was performed to identify relevant literature.  
Results. Pancreatoblastoma arises most frequently in the body and/or tail, or involves the entire pancreas. Ultrasonography, CT and MRI show variable imaging features, but should in most cases permit preoperative distinction of pancreatoblastoma from other tumours that occur in this region in infancy and childhood. Detection of metastases in the liver, lymph nodes and peritoneal cavity is not significantly better with any one of these three modalities.  
Conclusion. Preoperative imaging with US, CT and/or MRI will usually suggest a correct diagnosis of pancreatoblastoma. Contrary to previous reports, the tumour arises in the pancreatic head in a minority of cases.

Introduction  
Pancreatoblastoma [1] is a rare malignancy of the pancreas that occurs almost exclusively in children. The radiological features of pancreatoblastoma have been described in several case reports and one small series [2, 3]. We present the imaging findings in four new patients and discuss these in the context of other published work.

Patients and methods  
The medical records and imaging studies of four children with pancreatoblastoma were reviewed. The findings were reviewed by at least three radiologists, and the presence or absence of certain radiological signs was determined by consensus.

Results  
The age, sex, tumour size and location, biopsy procedures and outcome of the four patients are summarized.
in Table 1. There was no significant discrepancy in tumour location or size between any of the imaging methods used.

Three patients underwent US at diagnosis. In each case the tumour was approximately isochoic to liver. Two tumours showed a homogeneous echo pattern; the third contained rounded areas of fluid. Colour Doppler imaging in one patient showed mild tumour vascularity, encasement of the common hepatic artery and obliteration of the superior mesenteric vein. Fine calcification was identified in one patient.

In one case, US failed to detect a tiny (about 4 mm) solitary liver metastasis, which was later proven at open biopsy. In the other two cases, liver metastases were clearly shown by US. Nodal metastases and ascites were each demonstrated in two patients, and omental and peritoneal metastases and biliary dilatation each in one.

At CT, the primary tumour showed lower attenuation than liver in all cases, with mild heterogeneous contrast enhancement (Fig. 1). The tumour tissue was heterogeneous in three children, and in the fourth was homogeneous except for small areas of fluid density. Ascites (Figs. 2, 3) was present in three patients, and vascular encasement (Fig. 2), nodal metastases (Figs. 2, 3), and fine calcification (Fig. 1) were each shown in two.

Multiple hypodense liver metastases (with central necrosis in two patients) were identified in three cases (Figs. 1, 3, 4). In the fourth patient, there was a single tiny liver nodule. Omental and peritoneal metastases were present in two patients (Figs. 1, 3) and biliary dilatation (Fig. 2) in a single patient.

MRI was performed in two patients, showing heterogeneous tumours, hypointense to liver on T1-weighted (T1-W) images (Fig. 4) and predominantly hyperintense on T2-weighted (T2-W) images (Fig. 2c). One tumour contained multiple small fluid areas. Both showed mild patchy contrast enhancement, vascular encasement and ascites. In one patient, biliary dilatation and nodal metastases were present (Fig. 2d). MR cholangiography was helpful to clarify the level of biliary obstruction in this case (Fig. 2e). In the other there were multiple liver...