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Pancreatoblastoma with metastatic retro-peritoneal lymph node and PET/CT.

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Abstract

A previously healthy 4 year-old-female, presented with petechial rash and low platelet count. No other symptoms. On abdominal ultrasound a 4.7 cm heterogeneous mass was demonstrated anterior to the left kidney. An abdominal MRI subsequently performed demonstrated a heterogeneously enhancing mass at the same location extending to the pancreas and spleen. A surgical biopsy of the mass was obtained. Pathology reported a malignant epithelioid neoplasm consistent with pancreatoblastoma. The mass demonstrated intense FDG uptake on PET and an FDG avid retrocaval lymph node.

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FIGURE 1:

Abdominal ultrasound was performed on 4-year-old girl presenting with skin rash.¹⁻⁵ A heterogeneous 4.7 mass anterior to the left kidney was demonstrated (white asterisk in **A**). MRI of the abdomen (**B**) was subsequently performed, demonstrating within the retroperitoneal space a heterogeneously mass centrally colligated (black asterisk in **B**).

FIGURE 2:

A FDG-PET scan was performed to stage the disease⁶⁻⁹. The retroperitoneal mass was FDG avid (**A**) with central photopenia suggestive of necrosis (white asterisk in **A**). A retrocaval lymph node was FDG avid (white circle in **A**). FDG uptake was also noted along the pancreas (arrows in **B**) and at the splenic hilum abutting the spleen (arrowhead in **B**). The extent of the disease is well shown in **C**, where it is demonstrated that there is not other metastatic localization of disease.

FIGURE 3.

(image from biopsy 200x magnification, beta-catenin stain): Biopsy and subsequent tumor excision via a distal pancreatectomy, splenectomy and partial gastrectomy with pancreaticojejunostomy and retroperitoneal lymph node dissection were performed. Immunohistochemical staining revealed abnormal (nuclear) beta-catenin expression only in squamoid nests (shown in figure 3), supporting pancreatoblastoma diagnosis. The tumor cells also labeled with chymotrypsin, chromogranin, and synaptophysin (not shown) ⁹⁻¹³. Analysis of the surgical specimen demonstrated invasion of the portal vein and submucosa of the stomach. Among 14 retroperitoneal lymph nodes removed, only the one shown to be hypermetabolic on PET-CT revealed metastasis. Given the metastatic lymph node, patient underwent chemoradiation therapy ¹⁴⁻¹⁷.