

## Case Report

# Pancreatoblastoma with multiple peritoneal metastases: a rare entity with atypical presentation and review of literature

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### ABSTRACT

Pancreatoblastoma is a rare primary pancreatic neoplasm that predominantly affects young children and characterized by unique clinical features. Its biological behavior is aggressive with frequent local invasion and metastases. Only hundred cases have been reported in medical literature and its radiological features have been infrequently studied. We report the sonographic, contrast enhanced computed tomography findings of pancreatoblastoma in a 3 year old boy.

**Keywords:** Pancreatoblastoma, Hepatoblastoma, Alpha fetoprotein, Ultrasonography, Computed tomography

### INTRODUCTION

Pancreatoblastoma is a rare childhood malignancy which is most often located in the pancreatic head and body. It is often large in size and that can develop beyond the limits of the pancreatic gland and there by poses difficulty in deciphering the actual origin of tumor and may be confused with other more common childhood malignancies such as hepatoblastoma. It sends metastases to lymph nodes, liver, lungs and spleen. Our case had metastases to peritoneum.

### CASE REPORT

A 3-yr old boy was brought to our hospital with complaints of rapid distention of abdomen.

On inspection patient had bulging flanks. On palpation hepatic span is increased and fluid thrill was present which suggested gross ascites.

Gray scale ultrasound and contrast enhanced computed tomography was performed for further evaluation of the patient.

Gray scale ultrasound imaging findings revealed large, ill defined, solid cystic hyperechoic mass involving pancreatic head and body showing mild increase in vascularity (Figure 1). Gross ascites was involving pelvic and peritoneal cavities and multiple peritoneal metastatic deposits. Liver was enlarged with mildly prominent intra hepatic biliary radicles, however liver was free from any focal or diffuse mass lesion (Figure 2).

Axial contrast enhanced computed tomography imaging findings revealed large, ill-defined, lobulated solid cystic mass arising from head of pancreas. Post contrast scans reveal heterogeneous enhancement showing non enhancing necrotic areas (Figure 3). Anteriorly mass is causing antero lateral displacement of bowel loops. Laterally mass is seen abutting liver and extending till the hilum causing mild dilatation of central and peripheral

intrahepatic bile ducts (Figure 4). Posteriorly mass was extending till inferior vena cava and was causing compression of IVC with displacement towards left side. Medially mass was causing compressive dilatation of pancreatic duct and was displacing celiac artery laterally. Gross ascites and multiple peritoneal deposits were noted (Figure 5).



**Figure 1: Gray scale ultrasonography shows large, relatively solid hyperechoic mass with few cystic changes.**



**Figure 2: Sonogram reveals enlarged liver with prominent intra hepatic dilated radicles which indicates the reason behind confusing this mass with hepatoblastoma.**



**Figure 3: Axial contrast enhanced CT image shows large solid mass with few cystic areas involving head of pancreas causing dilatation of pancreatic duct.**



**Figure 4: Axial contrast enhanced CT image shows multiple hypodense enhancing peritoneal metastatic deposits.**



**Figure 5: Axial contrast enhanced CT image shows hepatomegaly with dilated intrahepatic biliary ducts. However, liver is free from any focal or diffuse mass lesion.**

## DISCUSSION

Pancreatoblastoma is a rare primary neoplasm of childhood, usually presenting in children ranging from 2-8 years old. Pancreatoblastoma was first described by Becker in 1957, which was termed as infantile pancreatic carcinoma.<sup>1</sup> Histologically tumor resembles foetal pancreatic tissue and hence the term pancreatoblastoma was termed by Horie.<sup>2</sup>

Pancreatoblastomas are composed of endocrine, acinar, ductal and islet cell elements.<sup>3,4</sup> By immuno histochemistry, the tumors exhibit positivity for pancreatic enzymes, endocrine markers and carcino embryonic antigens.<sup>4</sup>

Microscopically it is composed of variable elements of stroma and polygonal epithelial cells arranged in acinar or glandular pattern. The presence of Squamoid corpuscles, a loose aggregate of larger epitheloid cells of uncertain origin is a constant finding.

Pancreatoblastoma usually affects children ranging from one to eight years old with a predilection for males and Asians, but the entity also occurs rarely in adults.<sup>5-8</sup> At presentation most common complaints include abdominal pain and abdominal mass. Pancreatoblastoma is slow-growing, clinically occult, and quite large at the time of diagnosis.<sup>13</sup> Symptoms include early satiety, vomiting, constipation, and pain. Cushing syndrome and the syndrome of inappropriate antidiuretic hormone secretion have been reported.<sup>18</sup> Our case presented with jaundice and on examination had hepatomegaly and signs suggestive of gross ascites.

Differential diagnosis of pancreatoblastoma includes hepatoblastomas as both present with elevated alpha fetoprotein.<sup>9</sup> Pancreatoblastomas has several similarities with hepatoblastoma, both having associations with Beckwith-Wiedemann. Other differential diagnosis includes large intra- or retroperitoneal mass, such as a neuroblastoma, non-Hodgkin lymphoma, Wilms tumor.

Non-Hodgkin's lymphoma has hepato splenomegaly at presentation, and the affinity of the tumor for gallium helps to distinguish it from pancreatoblastoma.<sup>11</sup>

Since pancreatoblastoma has varied amount of cystic component, other pancreatic origin differentials include pseudocyst, pancreatic neoplasms and abscess.

Other serological markers associated with pancreatoblastoma include elevated lactate dehydrogenase, alpha -1 antitrypsin and Ca 19-9.<sup>10</sup>

The histopathologic features of pancreatoblastoma are well described in literature but very few series describe its imaging findings.

On gray scale ultrasonography pancreatoblastomas appear as well marginated, heterogeneous solid cystic masses with hyperechoic internal septations. Small punctuate calcifications may be present.<sup>11-14</sup> In our present case the tumor was very large and had heterogeneous solid cystic appearance causing mass effect and dilatation of intrahepatic biliary radicles. A provisional diagnosis of hepatoblastoma was made. Child also had gross ascites and peritoneal metastases. On contrast enhanced CT imaging origin of the tumor was from pancreas and displaced liver superiorly, hence diagnoses of pancreatoblastoma was made. Pancreatoblastomas rarely cause dilatation of biliary radicles owing to their soft consistency, however our case had this atypical finding.<sup>17</sup> The site of metastases is usually liver and lymphnodes. Other sites include lung and brain. Metastases to peritoneum are infrequently reported.<sup>3,12</sup>

In Montemarano et al. case series, in only half of the cases did imaging suggest pancreas as the organ of origin as these tumors were very large at the time of presentation.

Lee et al.<sup>14</sup> described pancreatoblastoma as partially or well-defined hypoattenuating solid cystic lesion with multiloculated elements and enhancing septae. Infiltrating margins are uncommonly reported.<sup>15</sup> Encasement of large arteries is also reported.<sup>12</sup>

Pancreatoblastomas are well marginated with low to intermediate signal intensity on T1WI and heterogenous signal intensity on T2WI.<sup>11,16-19</sup> Our patient did not undergo MRI due to financial constraints.

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## REFERENCES

1. Becker WF. Pancreatoduodenectomy for carcinoma of the pancreas of infant: report of a case. *Ann Surg.* 1957;145:864-70.
2. Horie A, Yano Y, Kotoo Y, Miwa A. Morphogenesis of pancreatoblastoma, infantile carcinoma of the pancreas: report of two cases. *Cancer.* 1977;39:247-54.
3. Montemarano H, Lonergan G, Bulas D, Selby D. Pancreatoblastoma: imaging findings in 10 patients and review of the literature. *Radiology.* 2000;214:476-82.
4. Klimstra DS, Wenig BM, Adair CF, Hefess CS. Pancreatoblastoma: a clinicopathologic study and review of literature. *Am J Surg Pathol.* 1995;12:1371-89.
5. Chung EM, Travis MD, Conran RM. Pancreatic tumors in children: radiologic-pathologic correlation. *Radiographics.* 2006;26:1211-38.

6. Kloppel G, Maillet B. Classification and staging of pancreatic nonendocrine tumors. *Radiol Clin North Am.* 1989;27:105-19.
7. Saif MW. Pancreatoblastoma. *JOP J Pancreas.* 2007;8:55-63.
8. Shorter N, Glick R, Klimstra D, Brennan MF, Laquaglia MP. Malignant pancreatic tumors in childhood and adolescence: the Memorial Sloan-Kettering experience, 1967 to present. *J Pediatr Surg.* 2002;37:887-92.
9. Defachelles AS, Martin De Lassalle E, Boutard P, Nelken B, Schneider P, Patte C. Pancreatoblastoma in childhood: clinical course and therapeutic management of seven patients. *Med Pediatr Oncol.* 2001;37:47-52.
10. Du E, Katz M, Weidner N, Yoder S, Moosa AR, Shabaik A. Ampullary Pancreatoblastoma in an elderly patient: a case report and review of the literature. *Arch Pathol Lab Med.* 2003;127:1501-5.
11. Cohen MD. Imaging of children with cancer. In: Cohen MD, eds. *A Book.* 3rd ed. St. Louis, Mo: Mosby-Year Book; 1992; 20-24, 52-62, 89-106, 134-156.
12. Gupta AK, Mitra DK, Berry M, Dinda AK, Bhatnagar V. Sonography and CT of pancreatoblastoma in children. *AJR Am J Roentgenol.* 2000;174(6):1639-41.
13. Passmore SJ, Berry PJ, Oakhill A. Recurrent pancreatoblastoma with inappropriate adrenocorticotrophic hormone secretion. *Arch Dis Child.* 1988;63:1494-6.
14. Roebuck DJ, Yuen MK, Wong YC, Shing MK, Lee CW, Li CK. Imaging features of pancreatoblastoma. *Pediatr Radiol.* 2001;31(7):501-6.
15. Lee JY, Kim IO, Kim WS, Kim CW, Yeon KM. CT and US findings of pancreatoblastoma. *J Comput Assist Tomogr.* 1996;20:370-4.
16. Immanura A, Nakagawa A, Okuno M, Takai S, Komada H, Kwon AH, et al. Pabcreatoblastoma in an adolescent girl: case report and review of 26 Japanese cases. *Eur J Surg.* 1998;164(4):309-12.
17. Stephenson CA, Kletzel M, Seibert JJ, Glasier CM. Pancreatoblastoma: MR appearance. *J Comput Assist Tomogr.* 1990;14:492-3.
18. Mergo PJ, Helmberger TK, Buetow PC, Helmberger RC, Ros PR. Pancreatic neoplasms: MR imaging and pathologic correlation. *Radiographics.* 1997;17:281-301.
19. Lumkin B, Anderson MW, Ablin DS, McGahan JP. CT, MRI and color Doppler correlation of pancreatoblastoma: a case report. *Pediatr Radiol.* 1993;23:61-2.

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