

PANCREATOBLASTOMA- A CASE REPORT

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CASE PRESENTATION

A 3-year-old male child referred to the radiology department with complaints as explained by parents - loss of appetite & loss of body weight since 6 months, feeling of mass in upper abdomen since 1 month, and occasional vomiting since 1 month.

On local examination, a firm non-tender mass was palpable in epigastric region and left hypochondriac region.

DIFFERENTIAL DIAGNOSES

- 1) Neuroblastoma
- 2) Wilms Tumor
- 3) Hepatoblastoma and other primary liver tumours.
- 4) Non-Hodgkin's Lymphoma-especially Burkitt's lymphoma.

USG Findings-A large 10 x 8 cm well defined, predominantly hypoechoic soft tissue mass with few areas of anechogenicity noted in the left hypochondrium and epigastric region, probably arising from the pancreatic body and tail. The lesion was taking minimal flow on colour Doppler. The lesion was causing mass effect on the spleen, left kidney and stomach. Rest of the abdomen appeared normal.



Figure 1. USG Image Demonstrates a Large 10 x 8 cm Well Defined, Soft Tissue Density Mass with Cystic Areas arising from Body and Tail of Pancreas

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CECT Abdomen Findings

On Plain CT- A large well defined heterogeneous soft tissue density mass with solid and cystic components noted arising from the body and tail region of the pancreas, there was no calcification noted.

On Post Contrast Imaging- The lesion showed moderate heterogeneous enhancement with non-enhancing necrotic areas. Neither vascular invasion nor hepatic metastasis noted.

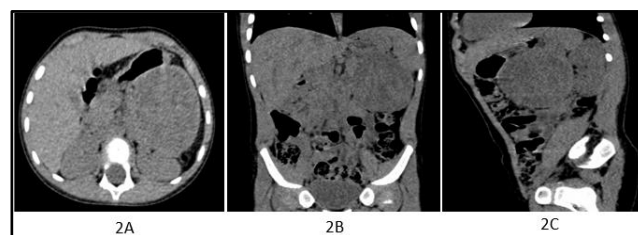


Figure (2A, 2B, 2C). Plain CT Images Demonstrates a Large Well Defined, Heterogeneous Soft Tissue Density Mass with Solid and Cystic Components in the Body and Tail Region of Pancreas

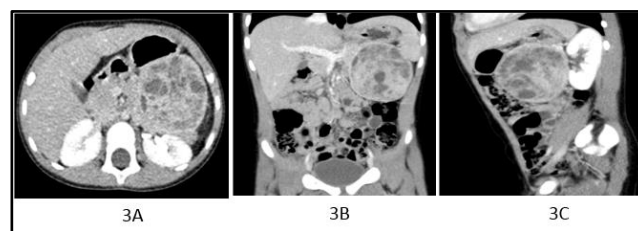


Figure (3A, 3B, 3C). Contrast CT Demonstrates Moderate Heterogeneous Enhancement of the Mass Lesion with Multiple Non-Enhancing Necrotic Areas, without Invasion of any Adjacent Structures/Vascular Invasion

CLINICAL DIAGNOSIS

Pancreatoblastoma.

DISCUSSION OF MANAGEMENT

The patient was taken for surgery by the paediatric surgery department.



Figure 4. Intra Operative Image Shows a Large Lobulated, Well Capsulated Lesion Noted in Body and Tail of Pancreas

Intra Operative Findings

A large well defined lobulated well circumscribed lesion with well-defined capsule noted in body and tail of pancreas. Which was resected and sent for histopathological examination.

PATHOLOGICAL DISCUSSION

Histopathological Findings

Encapsulated tumor composed of cellular stroma separating tumor into nests and trabeculae. The cells form solid structures resembling acini and short tubules. Cells lining the acini are small to medium sized round to polygonal with scanty eosinophilic cytoplasm. Eosinophilic secretions are noted within the lumen of some of the acini, focal capsular invasion and sub capsular vascular tumor emboli are also noted and diagnosis of pancreatoblastoma was made.

Paediatric Pancreatic tumours are quite rare in children, causing less than 0.2% of malignant paediatric deaths.¹ They are usually classified as exocrine or endocrine tumors. The most common exocrine pancreatic tumours in paediatric population are pancreatoblastoma and adenocarcinoma. Pancreatoblastoma arises from pancreatic acinar cells, these cells represent persistence of foetal anlage of the acinar cells.

Pancreatoblastoma also known as "infantile carcinoma of pancreas", is the most common pancreatic neoplasm of childhood.² This rare epithelial tumor, which is often misdiagnosed as neuroblastoma or hepatoblastoma,³ has a 2:1 male to female ratio.^{4,2} The mean age at diagnosis is 4 years, but it can manifest at any time from the newborn period to adulthood.⁴ More than half of reported cases are in Asians.^{5,6} Congenital cases of pancreatoblastoma have been described in association with Beckwith- Wiedemann syndrome, and these are predominantly cystic in nature.^{7,8}

Patients usually present with abdominal distension or a large, palpable abdominal mass. The mass may be associated with nonspecific symptoms, including failure to thrive, epigastric pain, anorexia, vomiting, diarrhoea and weight loss.² Obstructive jaundice may also be present. The serum α -fetoprotein level is elevated in 25% to 50% of patients, and the tumor may secrete adrenocorticotrophic hormone.^{4,5} Although usually located in the pancreatic

head,⁹ pancreatoblastoma can be located anywhere in the pancreas; it may be exophytic or may entirely replace the pancreas.² It tends to be a large mass with a diameter of upto 7 to 18 cm at the time of presentation. It is usually solitary, well defined, and surrounded by a fibrous capsule.^{9,2} Metastasis are most commonly seen in liver but may also occur in regional lymph nodes, the lungs and rarely in bone.⁵ Local invasion of the bowel and peritoneal cavity can also occur.² Metastatic disease is rare at the time of presentation. When metastatic disease is present the prognosis is typically poor.⁴

In general imaging demonstrates a solid mass with appearance suggestive of but not specific for pancreatoblastoma.¹⁰ Sonography demonstrates a well – demarcated, predominantly hypoechoic mass,⁵ which may have central areas of low echogenicity.²

CT demonstrates a focal mass with attenuation generally lower than that of liver. The mass may contain areas of low attenuation, and there is mild, heterogeneous contrast enhancement.⁵ The pancreatoblastoma usually is large and well defined, and it may be lobulated.³ Metastases to the Liver tend to be hypodense and may contain areas of central necrosis.⁵ Vascular encasement of the mesenteric vessels and inferior vena cava may develop, and calcifications can be present, which may make differentiation from neuroblastoma difficult.⁵

MR features of pancreatoblastoma are suggestive but not specific, with variable signal intensity on T1- weighted images and high signal intensity on T2-weighted images. The mass usually demonstrates enhancement after contrast administration.^{4,2} MR and CT can show direct invasion of adjacent structures, including the spleen, left kidney, left adrenal gland and omentum.⁵ Given the nonspecific imaging findings of pancreatoblastoma, the diagnosis is generally established by percutaneous biopsy.¹⁰

Treatment consists of surgical excision, with chemotherapy administered for metastatic disease.² Radiation therapy is used for local recurrence or incomplete resection.^{10,11} Although some patients are cured with excision alone, recurrence has been described in upto 60% of patients.¹¹

FINAL DIAGNOSIS

Pancreatoblastoma.

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