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Pancreatoblastoma in a 2.5-Year-Old Child: Imaging and Histopathologic Correlation in a Diagnostic Challenge

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Homi Bhabha Cancer Hospital (HBCH) & Mahamana Pandit Madan Mohan Malaviya Cancer Centre (MPMMCC)
Shreya Shukla
shreya@mpmmcc.tmc.gov.in
Homi Bhabha Cancer Hospital (HBCH) & Mahamana Pandit Madan Mohan Malaviya Cancer Centre (MPMMCC)
SN Singh
Homi Bhabha Cancer Hospital (HBCH) & Mahamana Pandit Madan Mohan Malaviya Cancer Centre (MPMMCC)
Soumitra Saha
Homi Bhabha Cancer Hospital (HBCH) & Mahamana Pandit Madan Mohan Malaviya Cancer Centre (MPMMCC)
Parichay Singh
Homi Bhabha Cancer Hospital (HBCH) & Mahamana Pandit Madan Mohan Malaviya Cancer Centre (MPMMCC)
Raghwesh Ranjan
Homi Bhabha Cancer Hospital (HBCH) & Mahamana Pandit Madan Mohan Malaviya Cancer Centre (MPMMCC)
Ajay Krishnan
Homi Bhabha Cancer Hospital (HBCH) & Mahamana Pandit Madan Mohan Malaviya Cancer Centre (MPMMCC)
Sambit Nanda
Homi Bhabha Cancer Hospital (HBCH) & Mahamana Pandit Madan Mohan Malaviya Cancer Centre (MPMMCC)

Case Report

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Abstract

Background:

Pancreatoblastoma is a rare malignant pancreatic tumor and the most common pancreatic malignancy in children. Its diagnosis is often difficult due to nonspecific presentation and overlap with other pediatric abdominal tumors.

Case Presentation:

A 2.5-year-old female presented with abdominal distension and intermittent pain. Examination revealed a large abdominal mass. Ultrasonography showed a heterogeneous solid-cystic lesion, and contrast-enhanced CT demonstrated a well-defined mass arising from the pancreatic body and tail with an enhancing solid component and no calcifications, displacing adjacent structures. PET-CT showed no metastases. Initial biopsy suggested neuroblastoma, and chemotherapy was started. However, following surgical excision, histopathological and immunohistochemical findings confirmed pancreatoblastoma.

Conclusion:

Pancreatoblastoma should be considered in young children presenting with large abdominal masses, particularly when imaging suggests pancreatic origin. This case highlights the risk of misdiagnosis and the importance of correlating imaging with histopathology for accurate diagnosis and management.

Introduction

Pancreatoblastoma is a rare malignant epithelial tumor of the pancreas, primarily affecting young children. It represents the most common pancreatic malignancy in the paediatric population, though only a limited number of cases have been reported worldwide. The median age at diagnosis is approximately four years, with a slight male predominance.[1,2] While the tumour typically occurs in early childhood, rare cases have been documented in adolescents and adults.

Clinically, patients often present with an abdominal mass and pain, and serum alpha-fetoprotein (AFP) levels are frequently elevated, reflecting tumor activity. [2]

Histologically, pancreatoblastoma exhibits acinar differentiation with characteristic squamoid morules and expression of pancreatic enzymes such as trypsin and chymotrypsin.[2,5] Radiologically, the tumor usually appears as a heterogeneous mass with solid and cystic or necrotic components, and calcifications may occasionally be seen. [7]

Locally, it may invade adjacent organs or encase major vessels, with the liver, lymph nodes, and lungs being common sites of metastasis.

Given its rarity, standardized diagnostic and therapeutic guidelines are lacking, and management is largely based on individual case experience.

We report a rare case of pancreatoblastoma in a 2.5 year old female child highlighting its imaging characteristics, histopathological features, and management approach.

Case report

A 2.5-year-old female child presented after her mother noticed abdominal distension for the past 2 3 days. The child had been experiencing intermittent abdominal pain and increased irritability for the preceding 2–3 months. There was no history of fever, vomiting, altered bowel or bladder habits, or any other swelling. Developmental milestones were age-appropriate, and there was no family history of malignancy.

On local examination, a firm, non-mobile mass was palpable in the left hypochondrium, extending cranio-caudally into the left lumbar region and transversely from the mid-axillary line to the epigastric area, measuring approximately 24 × 19 cm. The mass did not move with respiration. Ultrasonography (USG) demonstrated a large solid-cystic mass with internal vascularity, without calcifications {Figure 1}.

Non contrast and contrast-enhanced CT {Figure 2(a-d)} demonstrated a well-circumscribed, large solid-cystic mass in the left abdomen, extending from the left hypochondrium to the left lumbar region and crossing the midline. The mass contained a heterogeneously enhancing, eccentric, irregular solid component, with no evidence of calcifications. The lesion was along pancreatic body and tail causing posterior displacement of splenic vein. Positron emission tomography–computed tomography (PET-CT) showed no distant metastases.

The patient initially underwent a biopsy, and histopathological examination suggested a diagnosis of infantile neuroblastoma. Tumor cells were diffusely positive for synaptophysin and chromogranin, focally positive for CD99, diffusely positive for NKX2.2, and very focally and weakly positive for Sal-like protein-4 (SALL4), while negative for Glypican 3 and HepPar-1. Based on this provisional diagnosis, the patient was started on a regimen of Vincristine, Adriamycin, and Cyclophosphamide.

Subsequently, the patient underwent exploratory laparotomy, and the lesion was completely excised. Final histopathology revealed tumor cells that were positive for NKX2.2 and chromogranin, weakly and focally positive for synaptophysin, and negative for SALL4, Glypican 3, GATA3, and Cytokeratin 19. Occasional cells showed positivity for

Insulinoma-associated protein 1 (INSM1). Strong NKX2.2 positivity raised consideration for adamantinoma-like Ewing sarcoma; however, molecular testing did not identify an EWSR1 gene rearrangement. Considering the histomorphological and immunohistochemically features in the context of clinical presentation, the findings were most consistent with pancreatoblastoma

Timeline

Date / Period	Clinical Event
Jan–Feb 2024	Abdominal distension and intermittent pain for 2–3 months
07–08/02/2024	Initial evaluation; CECT showed large left-sided cystic abdominal mass
14/02/2024	PET-CT: no metastases; empirical VAC chemotherapy started
21/02/2024	Biopsy suggestive of neuroblastoma.
23/02/2024–02/03/2024	Hospital admission for supportive care and further workup
04/03/2024	Exploratory laparotomy with complete excision of mass
March–April 2024	Histopathological uncertainty; chemotherapy continued; tumor board discussion
May 2024	Final diagnosis confirmed as pancreatoblastoma; treatment completed
2024 (follow-up)	R0 resection status; patient clinically stable
2025	Surveillance imaging showed liver lesions; FNAC negative; AFP normal
Jan 2026	Latest follow-up: patient clinically stable

Discussion

Pancreatoblastoma (PB) is rare malignant epithelial neoplasm of the pancreas, representing less than 1% of all pancreatic tumours and primarily affecting the pediatric population. Since its first description by Becker in 1957, only a few hundred cases have been documented in the literature, reflecting its rarity and diagnostic challenge.[3] The tumor most frequently affects young children, with a mean age of presentation around five years, and shows a slight male or Asian predominance. [1,2] However, cases have also been described in adolescents and adults, indicating a broad clinical spectrum.

Histogenetically, PB is thought to arise from multipotent embryonic pancreatic cells that retain the potential for both endocrine and exocrine differentiation. Horie et al. proposed a

classification of PB into right- and left-sided variants, corresponding to origin from the ventral and dorsal pancreatic buds, respectively.[4] Right-sided tumors typically lack calcification and islet cell components, are well encapsulated, and carry a more favourable prognosis, while left-sided lesions often contain calcifications and islet cells, lack encapsulation, and show a more aggressive course. The present case displayed features of right sided tumor, showing a well-defined capsule and absence of calcification.

At the molecular level, several studies have demonstrated alterations in the APC/β-catenin signalling pathway, leading to aberrant nuclear accumulation of β-catenin in tumor cells, while p53 and K-ras mutations are usually absent. [5] Loss of heterozygosity on chromosome 11p has also been reported, supporting the association of PB with syndromes such as Beckwith–Wiedemann and familial adenomatous polyposis. These findings strengthen the concept that PB shares pathogenetic pathways with other embryonal tumors like hepatoblastoma and acinar cell carcinoma. Immunohistochemically, PBs express cytokeratin, epithelial membrane antigen, and pancreatic exocrine enzymes such as trypsin and chymotrypsin, while β-catenin positivity and E-cadherin loss may be observed in solid areas. [5] In our case, positivity for NKX2.2 and chromogranin helped in narrowing down differentials and on correlating with morphological features a diagnosis of Pancreatoblastoma was made.

The mass can exert a mass effect on adjacent structures, with characteristic displacement patterns that help localize the lesion to the pancreas[6]—for example, inward and anterior displacement of the superior mesenteric artery (SMA) and vein (SMV) with outward bowing of the duodenum in head lesions, or posterior displacement of the splenic artery and pancreas in body and tail lesions. Local invasion into adjacent organs such as the spleen, duodenum, or major vessels may occasionally be observed; however, distant metastases are uncommon at presentation in children.

Radiological differentials for large abdominal masses in children include hepatoblastoma (arising from the liver and supplied by hepatic arteries), retroperitoneal neuroblastoma (originating from the sympathetic chain, often encasing vessels and displacing the splenic artery anteriorly), lymphoma (typically involving multiple organs without calcification or necrosis), solid pseudopapillary neoplasm (usually in adolescent females, more homogeneous, rarely calcified), and acinar cell carcinoma (older children/adults, homogeneous enhancement, rare calcifications). Recognition of imaging characteristics—such as heterogeneous solid-cystic composition, calcifications, enhancement patterns, and vascular displacement—on CT, together with clinical and laboratory findings including age and serum alpha-fetoprotein levels, is crucial for suggesting pancreatic origin and narrowing the differential diagnosis. Careful integration of these features supports pancreatoblastoma as the most likely diagnosis in paediatric patients.[7]

Treatment of PB relies primarily on complete surgical resection, which remains the most significant prognostic factor. Studies by Dhebri et al. and Bien et al. indicate 5-year survival rates of 30–65% following total resection, compared with markedly poorer outcomes in patients with unresectable or metastatic disease at diagnosis. The role of chemotherapy is supportive, particularly for unresectable, recurrent, or metastatic lesions. Regimens incorporating agents such as cisplatin, etoposide, cyclophosphamide, vincristine, and doxorubicin have demonstrated partial efficacy, although there is no universally accepted protocol. Neoadjuvant chemotherapy may occasionally reduce tumor bulk to permit subsequent surgical removal. In extensive hepatic metastasis, liver transplantation has been proposed as a salvage option in selected cases.[8]

Given its rarity, the biological behaviour of pancreatoblastoma (PB) remains incompletely understood. Larger multicentre studies incorporating molecular profiling are needed to identify prognostic biomarkers, evaluate targeted therapies, and establish standardized chemotherapy protocols. Early recognition and aggressive multimodal management are essential to optimize survival outcomes. Our case illustrates the clinical, radiological and pathological features of PB and emphasizes the importance of continued accumulation of imaging data to refine diagnostic and therapeutic strategies.

Declarations

Ethics approval and consent to participate

-Ethical approval was not required for this case as per institutional policy. Informed consent was taken from patient’s guardians.

Consent for publication-

Written informed consent for publication was obtained from patient’s guardians.

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Author Contribution

The original draft was prepared by SS. Review and editing were carried out by SS, L, SNS, SS, PS, RR, AK, SN. All authors have read and approved the final version of the manuscript.

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Figures

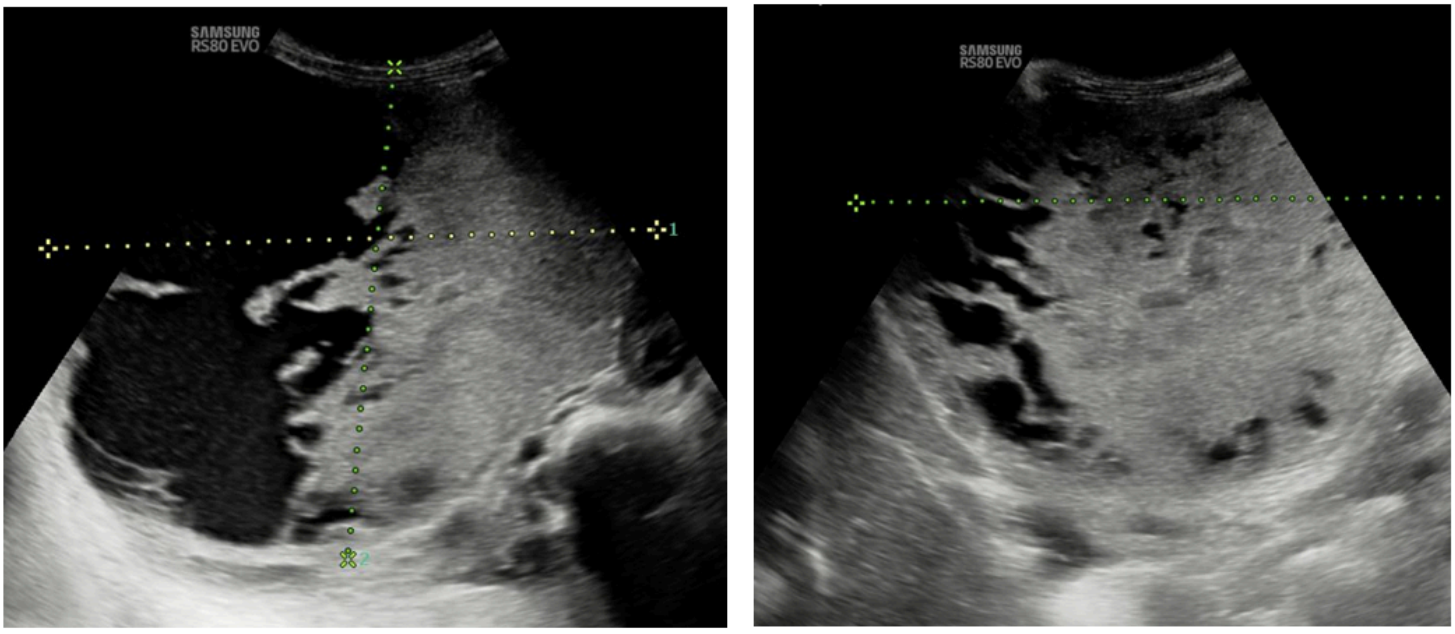


Figure 1

Ultrasound images show large heterogeneous solid-cystic mass in left hypochondrium

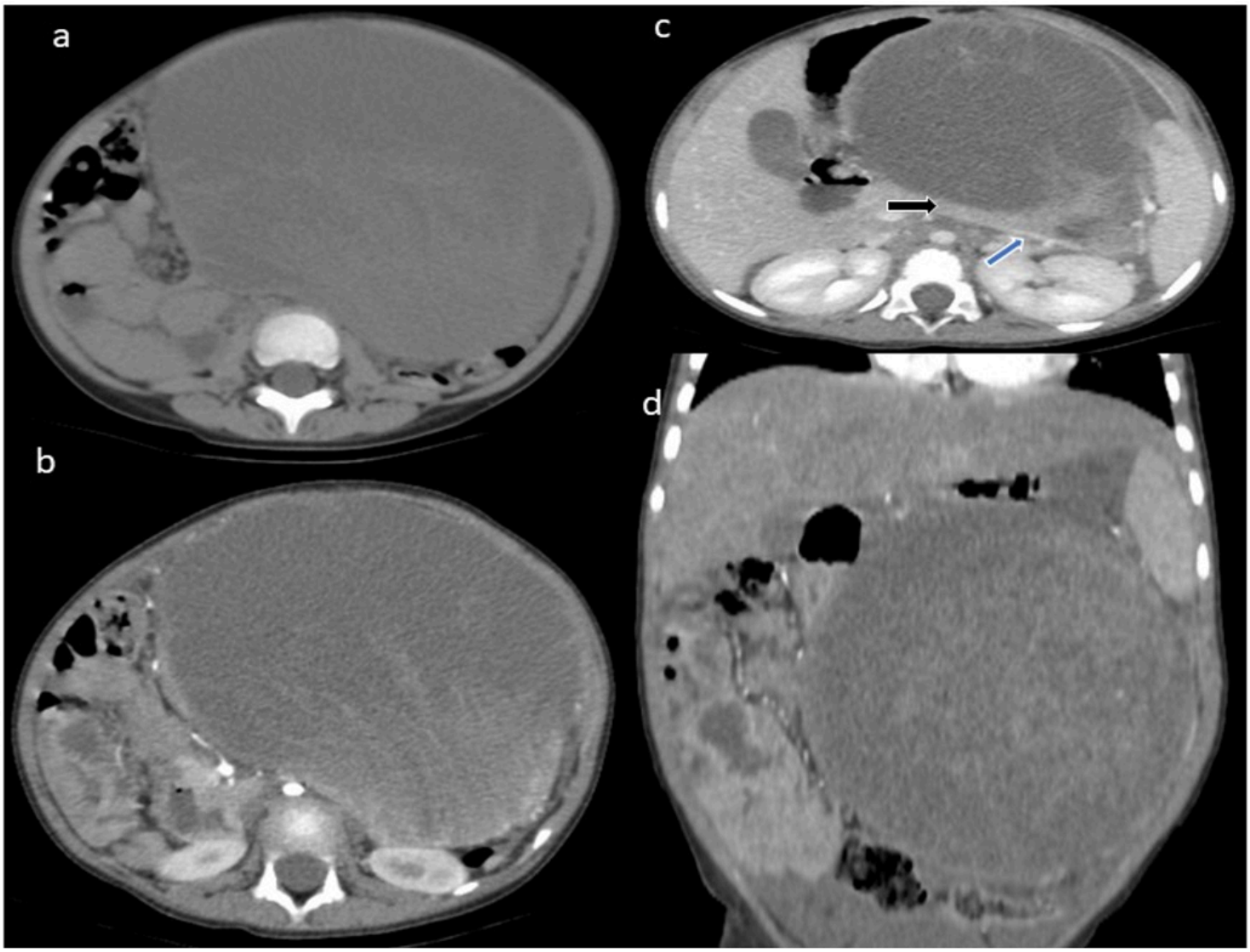


Figure 2

(a–d): NCCT and post contrast images of the abdominal mass. (a) Axial NCCT image shows a large heterogeneous mass on left side. (b) Post contrast axial image shows heterogeneously enhancing peripheral solid component within mass. (c) Post contrast axial image shows relation with pancreatic body and tail (Black arrow) and posterior displacement of splenic vein (Blue arrow). (d) Post contrast Coronal images show extent of the lesion from left hypochondrium till iliac region.

Supplementary Files

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- CAREchecklist.pdf