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## CASE REPORT

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# Two Children with Pancreatoblastomas

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### ABSTRACT

*Pancreatoblastoma is a rare primary pancreatic neoplasm occurring in children. This report describes two paediatric patients with pancreatoblastomas with characteristic sonographic and computed tomographic features. Characteristic clinical and imaging features, and the differential diagnosis are discussed.*

*Key Words: Child; Pancreatic neoplasms; Tomography, X-ray computed; Ultrasonography*

## 中文摘要

### 兩名胰腺母細胞瘤小兒患者

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胰腺母細胞瘤是一種罕見的發生在兒童的原發性胰腺惡性腫瘤。本文報告有超聲波及電腦斷層造影特徵的兩名胰腺母細胞瘤小兒患者。並討論此病的臨床、影像學特徵及其鑒別診斷。

### INTRODUCTION

Pancreatoblastoma is a rare primary neoplasm of childhood, first described in a 15-month-old boy as “infantile adenocarcinoma of the pancreas” by Becker<sup>1</sup> in 1957. In 1977, Horie et al<sup>2</sup> advocated using the term pancreatoblastoma for this tumour, based on its histological resemblance to incompletely differentiated acini of fetal origin at approximately 7 weeks of gestation. Since then, approximately 100 cases of pancreatoblastoma have been reported in the medical literature.

Although the tumour usually affects children aged 1 to 8 years, rare cases have been reported in neonates and adults.<sup>3,4</sup> There is a male predominance ranging from 1.3:1 to 2.7:1.<sup>5-7</sup> More than half of the reported cases were in Asians.<sup>1</sup> It is unclear whether the apparent high incidence in Asians reflects a true

difference in incidence or greater interest in the entity and consequently more comprehensive reporting of pancreatoblastoma in Far-Eastern countries.<sup>8</sup>

Elevated  $\alpha$ -fetoprotein level is the most common serological marker, noted in up to one-third of paediatric cases.<sup>3</sup> Elevated lactate dehydrogenase,  $\alpha$ -1 antitrypsin, and CA 19-9 have also been associated with pancreatoblastoma.<sup>3,9</sup> Although most cases are sporadic, congenital cases of pancreatoblastoma have been described in association with the Beckwith-Wiedemann syndrome, and these are predominantly cystic in nature.<sup>6,10</sup> Most commonly, patients present with an asymptomatic, large abdominal mass.<sup>4,11</sup> Symptoms that may ensue are usually non-specific, and include abdominal pain, weight loss, anorexia, diarrhoea, and vomiting.<sup>4,6,11</sup>

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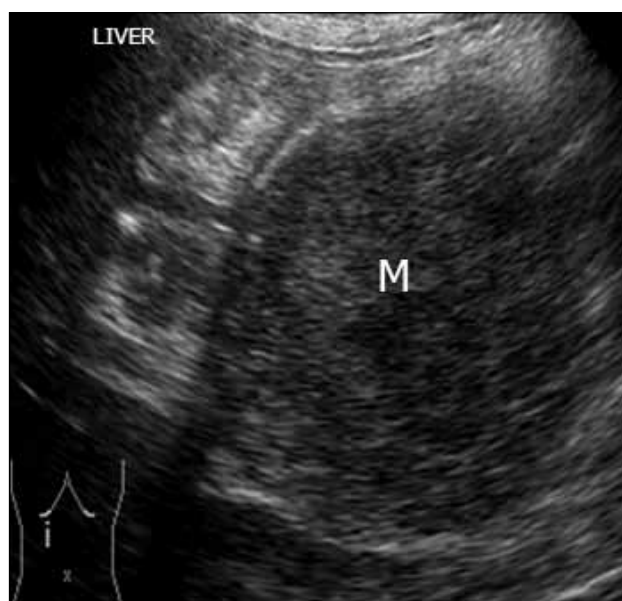
### Case 1

A 9-year-old girl presented with recent-onset epigastric pain and discomfort in February 2004. A fixed hard mass was felt in the epigastric region. An initial ultrasound revealed a heterogeneously hypoechoic mass with an epicentre at the head of pancreas (Figure 1). Subsequent computed tomographic (CT) examination showed a well-encapsulated mass arising from the head of pancreas with peripheral solid and central necrotic components. The right kidney, right renal vein, and inferior vena cava were displaced posteriorly (Figure 2). There was no invasion into the surrounding structures nor were any systemic metastases identified. Based on imaging and clinical features, a diagnosis of pancreatoblastoma was suggested.

The patient underwent complete resection of the tumour, and the diagnosis was proven on histology. The girl is in disease remission, without any suggestion of recurrence based on postoperative 2-year follow-up imaging.

### Case 2

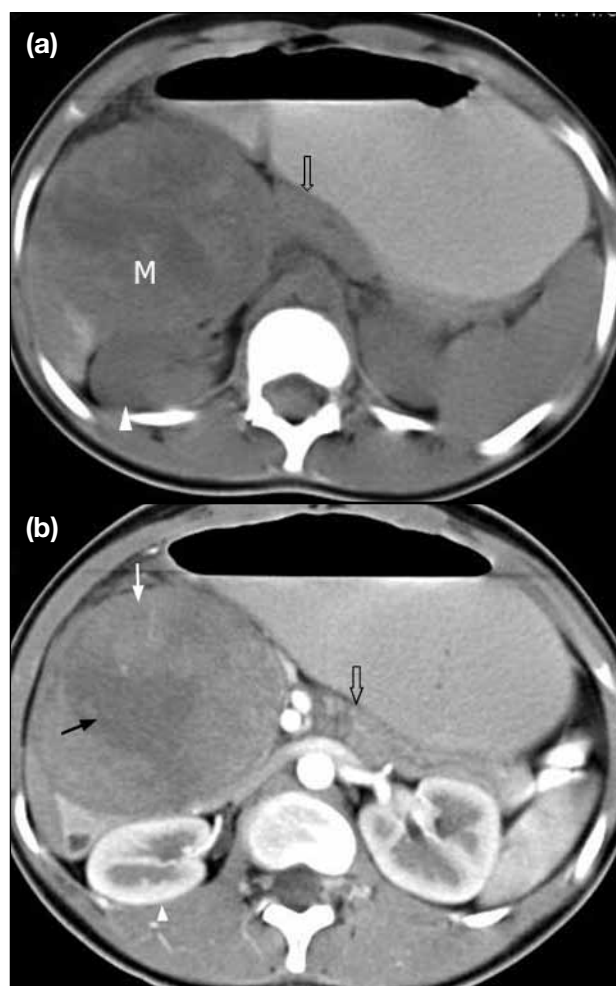
An 11-year-old girl presented in May 2004 with pain and discomfort in the epigastric region, a lump in abdomen, and vomiting after meals. There was a firm-hard swelling in epigastric region. Ultrasound showed a hypoechoic mass at the splenic bed. The splenic vessels were displaced inferolaterally (Figure 3), but its exact



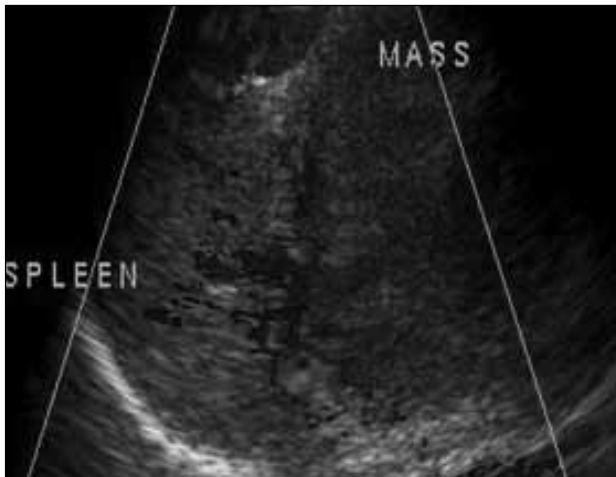
**Figure 1.** Grey-scale ultrasound shows a large roundish heterogeneously hypoechoic mass (M) in the head of pancreas.

origin of the mass was difficult to establish. However, CT revealed the typical beak sign confirming its origin from the distal body and pancreatic tail. Peripheral enhancing solid and central non-enhancing central components were detected after intravenous contrast administration (Figure 4). The mass was well localised and there was no evidence to indicate local spread or systemic metastases.

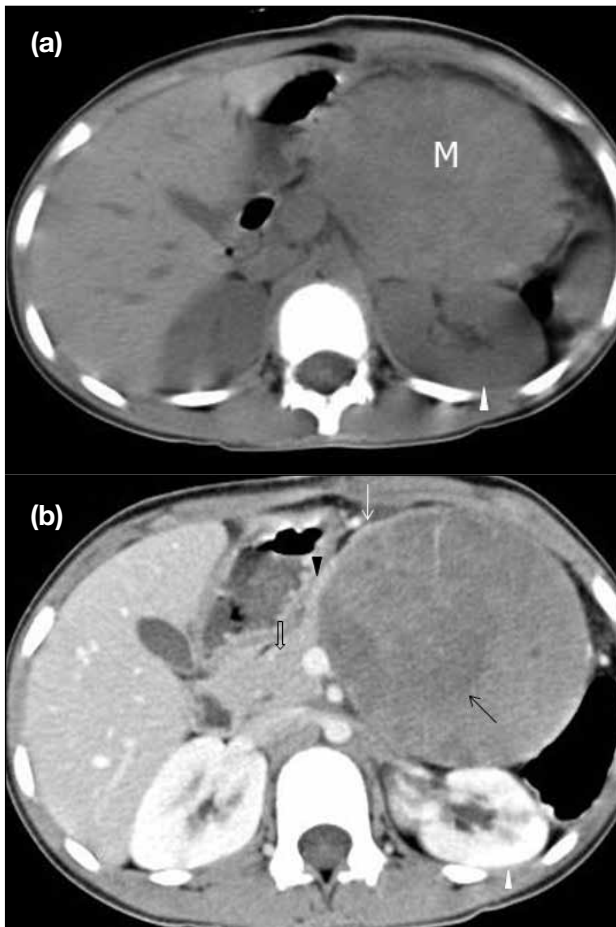
As in the first case, the patient underwent complete surgical resection and histopathology confirmed it was a pancreatoblastoma. Follow-up imaging at 6 months and 2 years after surgery was unremarkable, with no disease recurrence.



**Figure 2.** (a) Non-enhanced and (b) contrast-enhanced axial computed tomographic images showing an encapsulated mass (M) arising from the head of pancreas. Note the enhancing peripheral solid (white arrow) and non-enhancing central necrotic components (black arrow). The right kidney (white arrowhead) is displaced posteriorly. The normal pancreas is shown (open arrow). There was no invasion into the surrounding viscera.



**Figure 3.** A Doppler transverse image at the splenic bed shows a large hypoechoic mass displacing the splenic vessels inferolaterally.



**Figure 4.** (a) Non-enhanced, and (b) contrast-enhanced axial computed tomographic images showing a large mass (M) in the distal body and tail of pancreas as evident by beak sign (black arrowhead). The mass is well encapsulated (white arrow) with predominant solid enhancing components. Note the patchy areas of non-enhancing central necrosis (black arrow). The left kidney (white arrowhead) is displaced posteriorly. The normal pancreas is shown (open arrow).

## DISCUSSION

Pancreatoblastomas are usually large, solitary masses; ranging in diameter from 1.5 to 20 cm, with a mean of 10.6 cm.<sup>6</sup> In approximately half of the patients, it arises in the head of the pancreas.<sup>4,5</sup> The tumour is a well-defined or partially circumscribed, solid mass with lobulated margins.<sup>5,6,12</sup> Although the pathological features of pancreatoblastoma are well described in the literature, few reports describe its radiological features.

The mass can be so large at presentation that determination of origin becomes difficult. In the series of Montemarano et al,<sup>4</sup> in only half of the cases did the imaging suggest the pancreas as the organ of origin. These large tumours typically compress surrounding structures without radiological evidence of invading them, although local invasion may be evident at surgical resection.<sup>4,11</sup> Dilatation of the biliary tree is uncommon, most probably due to the soft consistency of the tumour.<sup>4</sup> Encasement of large arteries has been reported.<sup>7,11</sup>

At sonography, the majority of pancreatoblastomas appear to be well-circumscribed heterogeneous masses, with solid and cystic components<sup>4,7</sup> as seen in our first case. Cystic structures are hypoechoic with hyperechoic internal septa.<sup>4,11,13</sup> Occasionally, a hypoechoic solid mass is encountered<sup>12</sup> as in our second case. The mass is most commonly well or partially circumscribed on CT images, although an infiltrative margin is uncommonly noted.<sup>4</sup> The tumour is often smooth and may be multilobulated.<sup>13</sup> In the majority of cases, the mass is heterogeneous due to internal cystic areas, reflecting the regions of necrosis seen at pathological inspection. Both our patients showed similar CT features. Frequently, the tumour appears multiloculated with enhancing septa.<sup>4,8,13</sup> Small punctate, clustered, or curvilinear calcifications may be identified.<sup>4,13</sup> On magnetic resonance imaging (MRI), pancreatoblastomas are well margined with low-to-intermediate signal intensity on T1-weighted images and heterogeneous high signal intensity on T2-weighted images.<sup>4</sup> Low signal intensity on T1-weighted images corresponds to foci of necrosis.<sup>14</sup> Enhancement with contrast is a common feature seen with CT and MRI. No studies suggest significant differences in imaging findings between adult and paediatric patients. Invasion of adjacent organs and distant metastases may occur with pancreatoblastoma. If locally advanced, the tumour is poorly margined and invades the surrounding pancreas, peripancreatic tissues, and adjacent organs.<sup>6</sup> Biliary invasion has been

reported.<sup>4</sup> Vascular invasion is rare, although portal and mesenteric vein invasion has been reported. At presentation, metastases to the liver and abdominal lymph nodes are found in about 35% of the patients in some series.<sup>4,11</sup> Less commonly, metastases are noted in the lung and brain.<sup>4,12</sup> Rare cases of metastasis to the omentum, pelvic cul-de-sac, colon, spleen, kidney, and adrenal glands have been reported.<sup>4,6</sup>

Pancreatoblastoma should be considered a top differential diagnosis for primary pancreatic neoplasms in the paediatric population, because of its high frequency in this age-group. Predominantly cystic pancreatoblastomas can appear radiologically similar to solid-pseudopapillary tumours. Cystic pancreatoblastomas, however, tend to occur in newborn patients with Beckwith-Wiedemann syndrome (more commonly in boys),<sup>15</sup> whereas solid-pseudopapillary tumours tend to arise in adolescent girls and young women. Acinar cell carcinomas have overlapping radiological features. The 2 can be distinguished from each other based on the age of the patient—pancreatoblastomas occur in children aged 10 years or less, whereas acinar carcinomas occur almost exclusively in older patients.<sup>16</sup> A large size and infiltrative margins are known imaging features of these rare tumours.<sup>4</sup> The differential diagnosis for large masses with less distinct organ of origin remains wide. Such tumours frequently occurring in young children include neuroblastoma, hepatoblastoma, other primary liver, renal and adrenal tumours.<sup>16</sup> Fortunately, both of our patients had well-defined tumour margins and the pancreatic beak sign helped identify the organ of origin.

Surgical resection is the mainstay of treatment for localised tumours and in paediatric cases without metastases, chances of survival are excellent.<sup>5,6,10,12</sup> In one series, 37% of cases presented with metastatic disease, for which treatment was supplemented by chemotherapy and radiation.<sup>9</sup>

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