



### Pancreatic Tumors in Children Our experience

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#### Abstract :

Five children presented to us with pancreatic tumors over a period of 7 years. The presentation included abdominal pain, jaundice, a history of trivial trauma, vomiting, and recurrence of disease. Ultrasound, CT Scan, and MRI were useful in diagnosis. The possibility of pseudocyst of pancreas was entertained by the radiologist after MRI. Surgical extirpation is necessary for cure. Due to their low invasive potential, tumor enucleation and spleen preserving distal or central pancreatectomy were performed in 3 children. The proximity of the portal vein and involvement of the hepatic artery make surgery treacherous and exciting. Postoperative follow-up ranged from 10 months to 48 months.

**Keyword :** Pancreatic Tumors, Children

#### Introduction:

Pancreatic Tumors in children are a rare entity. Very few cases have been described in literature. 5 children presented to us over a period of 7 years from January 2006 to December 2012.

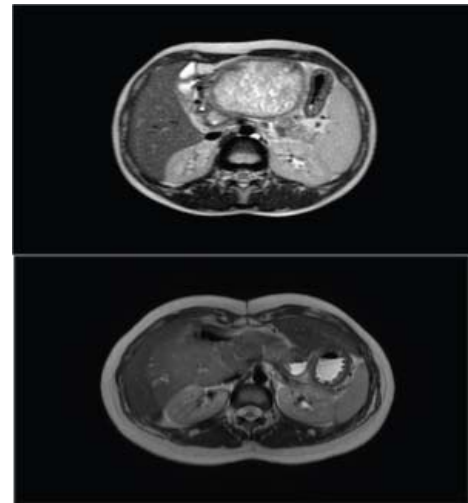
We present our experience from these patients. **Materials:** Five children presented to us over a 7 year period. There were two males and three females. They ranged in age at the time of presentation from 9 months to 14 years. The most common symptom at the time of presentation was pain, followed by a mass in the abdomen. 1 child presented with vomiting and one with jaundice. **Methods:** A retrospective study conducted from January 2006 to December 2012. All children below the age of 16 years, who presented to us with pancreatic tumors were included in this study. Their presentation, diagnosis, treatment and outcome are presented as such. **Case1:** A 9 month old boy presented to us with jaundice for a month prior to presentation. This child had a mass in the abdomen. CT Scan showed a large mass in the head of the pancreas which was encasing the portal vein. A laparotomy was done which revealed metastasis to the liver and a tumor which was occupying most of the head of the pancreas and encasing the portal vein. A biopsy was performed and the abdomen was closed.

The biopsy revealed a pancreatoblastoma. The child was given 6 cycles of adjuvant chemotherapy including Cisplatin and Doxorubicin. Repeat ultrasound and CT showed minimal response to chemotherapy. The child succumbed to the disease 10 months into treatment. **Case 2:** A 9 year old boy presented to us having had 3 laparotomies done in another institution. The child had a distal pancreatectomy with splenectomy initially. He had a recurrence for which 2 laparotomies were done in a space of 5 months. After the third laparotomy he developed a fecal fistula. A CT Scan done showed a large mass occupying most of the abdomen and the retroperitoneum. The child had sepsis and multi organ dysfunction syndrome. He was given palliative care in ICU and died due to sepsis after 10 days.

All these children had preop imaging with ultrasound and Computed tomography scan(CT) which revealed tumor in the central pancreas. A endoscopic ultrasound guided fine needle aspiration cytology was done for case 3. This showed features of solid pseudopapillary tumor. This tumor was subsequently excised using a distal pancreatectomy. No pre operative biopsy was done for Case 4 and 5. Surgery was done based on imaging studies. Biopsy was consistent with solid pseudopapillary tumor of the pancreas. No further treatment was given as the role of chemotherapy and radiotherapy is not defined. These children have been followed up for 24 – 48 months. Their post operative imaging with CT scan or MRI at 6 months has shown no recurrence of disease. Their USG imaging at 12 and 48 months has shown no recurrence of disease.

**Figure 1:** Pre (Picture 1) and post operative (Picture 2) MRI showing tumor in central Pancreas and no recurrence at 6 months

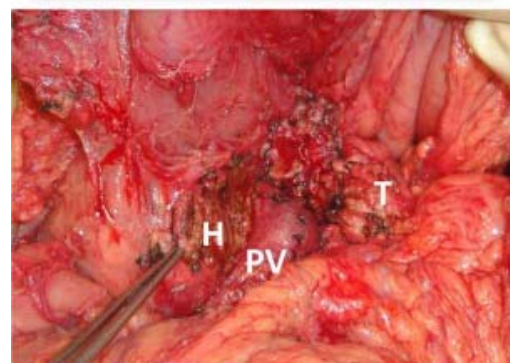
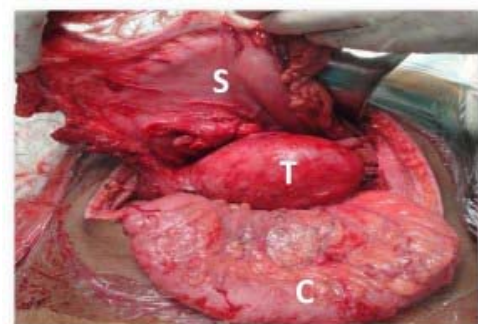
**Picture 1 Picture 2**



**Figure 2:** Peroperative photo (Picture 3) showing a large tumor (M) and its relation to stomach (S) and colon (C) and post excision (Picture 4) showing the Portal Vein (PV) head of Pancreas (H) and tail of pancreas (T).

**Picture 3 Picture 4**

**Discussion:**



Pancreatic tumors are rare entities in children<sup>(1)</sup>. The presentation in children is different, who unlike adults present with pain and mass instead of jaundice. Pancreatoblastomas are shown to have less mortality when they present without metastasis. Inflammatory myofibroblastic tumors (IMT) are rare, benign lesions. They mimic, clinically and radiologically, malignant tumors—especially sarcoma<sup>(2)</sup>. Solid Pseudopapillary Tumor (Frantz's tumor) occur predominantly in girls and young women. The tumors are well encapsulated and the cut surfaces, show characteristically solid and hemorrhagic-necrotic patterns. Ultrasonography and CT scan are the most useful tools for the diagnosis. The neoplasms usually behave like a very low-grade malignancy, so that complete removal is the treatment of choice for the tumor arising anywhere in the pancreas. These tumors also require close follow up with repeat imaging to rule out recurrence.<sup>(3)</sup> Unlike malignant pancreatic tumors in adults, tumors in children and adolescents usually are resectable, and long-term survival is likely. However, the risk of recurrence for pancreatoblastoma is high. The roles of chemotherapy and radiation remain undefined in most pancreatic tumors<sup>(4)</sup>. Survival was significantly greater for female patients and for those who had surgery<sup>(5)</sup>. The imaging appearance of each neoplasm reflects its pathologic features. Pancreatoblastoma is the most common pancreatic neoplasm in young children. At imaging, pancreatoblastomas are heterogeneous and often multilocular with hyperechoic and enhancing septa. Solid-pseudopapillary tumor occurs in adolescent girls. It is heterogeneous in internal architecture, with a mixture of solid and cystic hemorrhagic and necrotic elements. This tumor is distinguished by its fibrous capsule and hemorrhagic nature, which are best shown at magnetic resonance imaging as a dark rim on T1- or T2-weighted images and hyper-intense foci on

T1-weighted images, respectively. All pancreatic neoplasms in children are capable of producing metastases, usually to the liver and lymph nodes; however, on the whole, these tumors have a better clinical outcome than most pancreatic tumors in adults. Knowledge of the differential diagnosis of pancreatic masses in children and their relatively good prognosis may promote correct preoperative diagnosis and appropriate treatment<sup>(6)</sup>. Surgery remains the keystone of treatment for pancreatic tumors in pediatric age as in adults.

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**Table1 : Following is the description of cases 3,4 and 5**

Case	Age/Gender	Site	FNAC/ Biopsy	Surgery	Post Op Imaging
3	13year/female	Central Pancreas	Solid-pseudopapillary tumor	Spleen preserving Distal Pancreatectomy	No recurrence
4	13year/female	Central Pancreas	Solid-pseudopapillary Tumor	Central Pancreatectomy	No recurrence
5	13year/female	Central Pancreas	Solid-pseudopapillary Tumor	Central Pancreatectomy	No recurrence