

## CASE REPORT

# Two Cases of Adult Pancreatoblastoma: An Infrequent Differential Diagnosis

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### ABSTRACT

Pancreatoblastomas are extremely infrequent nonductal pancreatic tumors in adults; their behavior is usually more aggressive than in children. Surgical resection is the primary treatment, and other approaches are not universally accepted. Here, we present two cases operated at our institution. One case with a large tumor located at the body and tail of the pancreas; the other patient had the tumor located at the head of the pancreas, both were treated surgically and one case recurred during follow-up. Pancreatoblastomas should be considered as a differential diagnosis in adult population when clinical and radiologic features are not typical for adult pancreatic masses.

### INTRODUCTION

Pancreatoblastoma is a rare entity that represents <0.5% of all exocrine pancreatic neoplasms [1]. They usually appear in the first decade of life. Adult cases are extremely unusual and reported cases amount to 35, with the first one published in 1986 [2]. Clinical manifestations are usually unspecific, including the presence of mass; around 50% of the patients will experience abdominal pain [3]. Tumor markers are not useful as in pediatric populations, positivity for AFP and CA 19-9 is 6% [1]. Histological findings show ductal, mesenchymal, neuroendocrine and acinar differentiation, being the presence of squamous nest useful for diagnosis and distinction from acinar cell carcinomas [4]. The prognosis in these patients is poor compared to children population [5, 6]. Our purpose is to present two cases surgically treated at our institution and review current literature about this disease.

### CASE PRESENTATION

#### Case #1

A 41 year-old man was admitted to our institution due to abdominal pain and a large abdominal mass in the left upper quadrant. Upper digestive bleeding was characterized by melena. Weight loss of 10 kg in 4 months was noticed by the patient. His past medical history includes epilepsy for which he takes Carbamazepine. On physical examination, the patient looks with a good performance status but weight loss is evident; in the

abdomen a 25 cm tumor is palpated, the mass is apparently fixed to the retroperitoneum and is discretely tender on palpation. The CT scan shows a 30 cm mass that depends of the tail of the pancreas and extends to the splenic hilum (**Figure 1a**), portal phase shows a blood clot in the splenic vein (**Figure 1b**, arrow). The serum AFP levels were 4.410 ng/mL. CA 19-9 and CEA levels were normal. The patient had a corporocaudal pancreatectomy with splenectomy and left colectomy *en bloc* to ensure free surgical margins (**Figure 1c**). His post-operative period was uneventful and the patient was discharged on post-operative day 8. The pathology revealed a Pancreatoblastoma with infiltration to the serosa of the colon and lymphatic and vascular emboli, all surgical margins were negative due to extended resection, immunohistochemistry was positive for  $\beta$ -catenin, AFP, synaptophysin, and chromogranin; the Ki-67 index was 70%. The patient was sent for chemotherapy and Gemcitabine was started. AFP two months after surgery was 24.49 ng/mL, during follow-up (7 months after surgery) he developed Spinal Cord Compression Syndrome and Radiotherapy was given as palliation, after treatment the patient started second-line chemotherapy with Cisplatin-Doxorubicin. Last follow-up is 10 months after surgery and he is receiving chemotherapy with evidence of stable disease.

#### Case #2

An 18 year-old woman was admitted due to abdominal pain, jaundice and nausea and vomiting. Her past medical history is negative for diseases. On physical examination, the patient is jaundiced with a good performance status (ECOG 0). The abdominal findings show no abnormalities. The contrast-enhanced MRI with gadolinium shows a mass that depends of the uncinate process of the pancreas with associated dilation of the pancreatic and bile ducts (**Figure 2a**). Serum CA 19-9 levels were normal. The patient had a pancreaticoduodenectomy, during the procedure a

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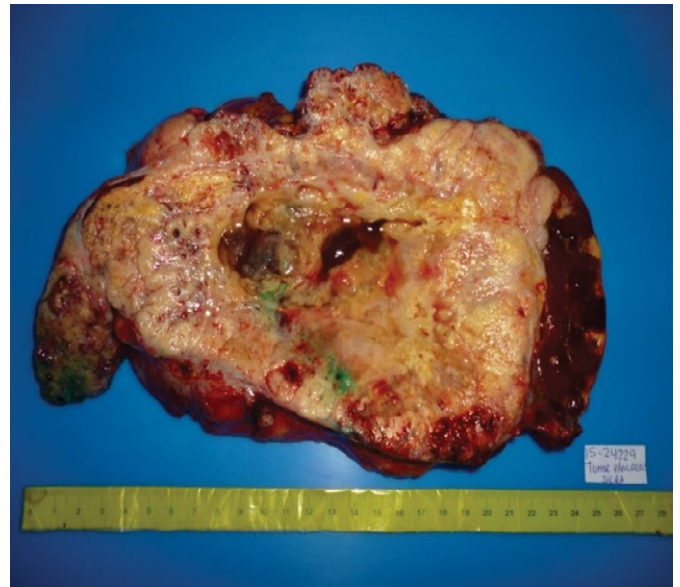
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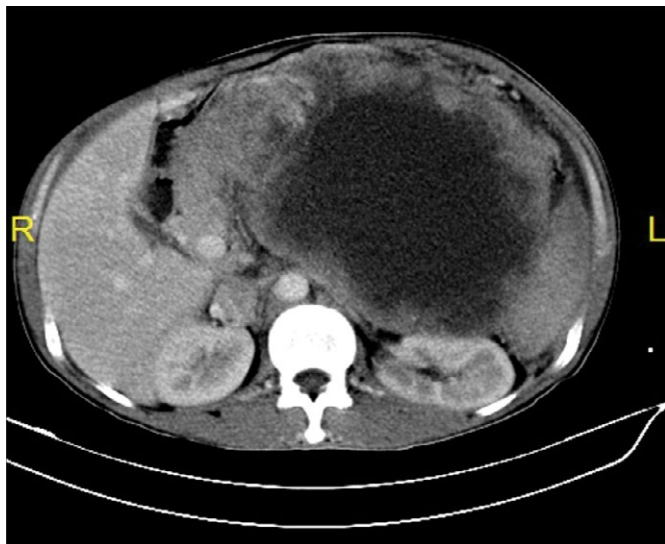
tumoral thrombus was found in the portal vein (**Figure 2b**) and required vascular resection and primary anastomosis (**Figure 2c**). Her post-operative period was uneventful and the patient was discharged on post-operative day 9. The pathology revealed a 4 cm in diameter Pancreatoblastoma with infiltration to the adjacent pancreatic tissue and distal bile duct (**Figure 3a**). No lymphatic or vascular embolus was seen; surgical margins were negative. Perineural infiltration was extensive. Immunohistochemistry was positive for  $\beta$ -catenin, pankeratin, synaptophysin, and chromogranin; a high number of mitoses were seen. Last follow-up is 15 months after surgery and the patient is recurrence free (**Figures 3b, 3c, 3d**).

### DISCUSSION

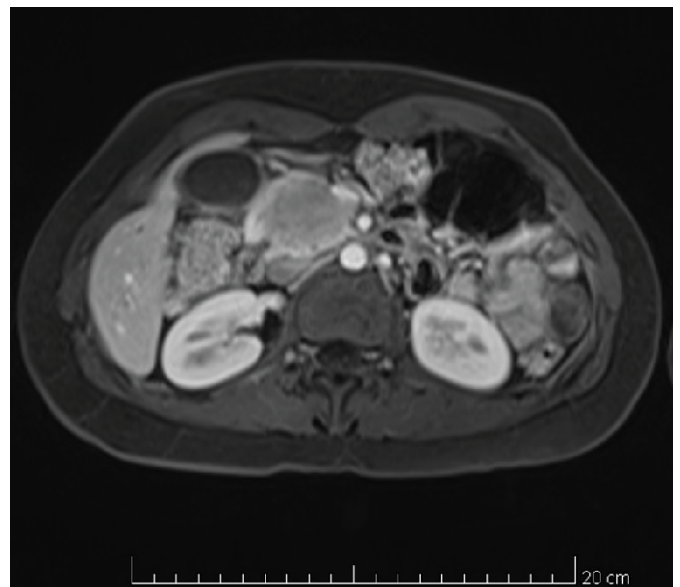
The term Pancreatoblastoma was first used by Horie in 1977 [7] and the first case in an adult was reported by



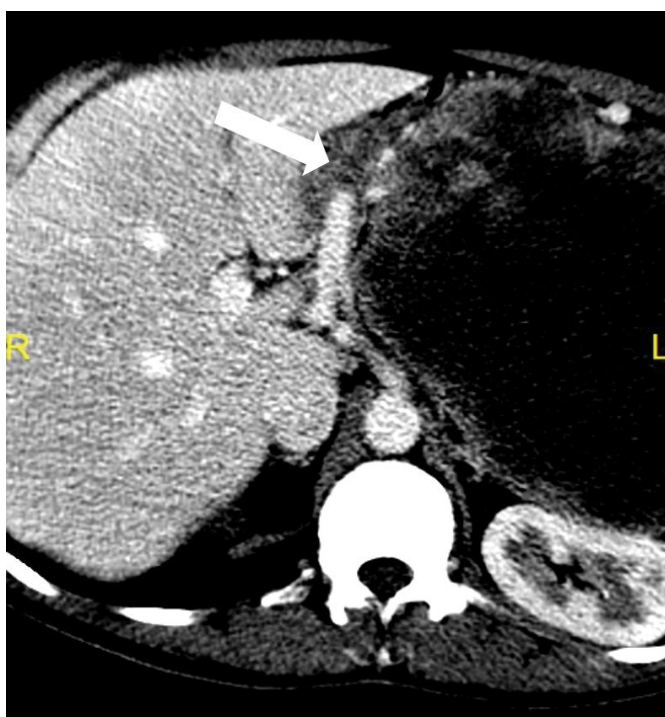
**Figure 1c.** Gross pathologic specimen shows a tumor depending of the tail of the pancreas with cystic and solid areas with lobulated and expansive appearance, partially encapsulated with extensive necrosis.



**Figure 1a.** Large mass with hypodense center that depends on the tail of the pancreas.



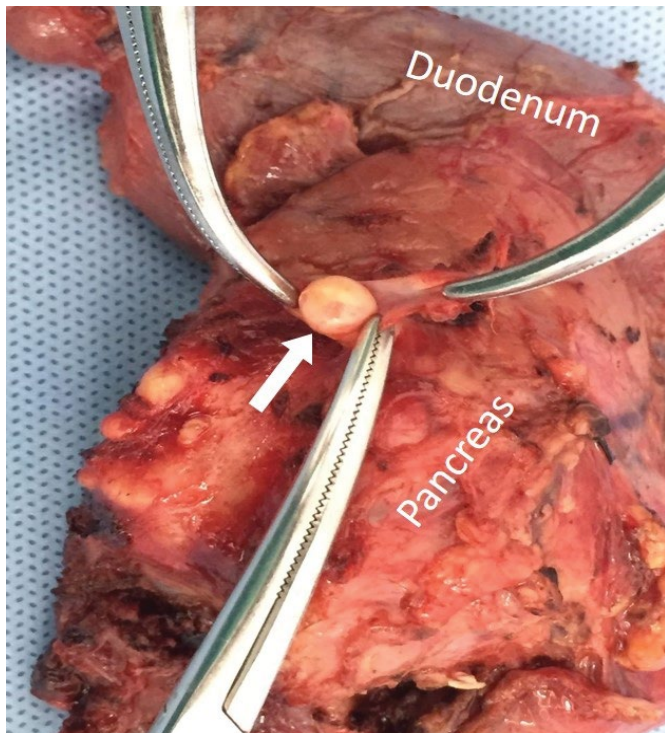
**Figure 2a.** MRI shows a pancreatic head and uncinate process mass that displaces the mesenteric vein in anterior direction, the mesenteric artery is intact.



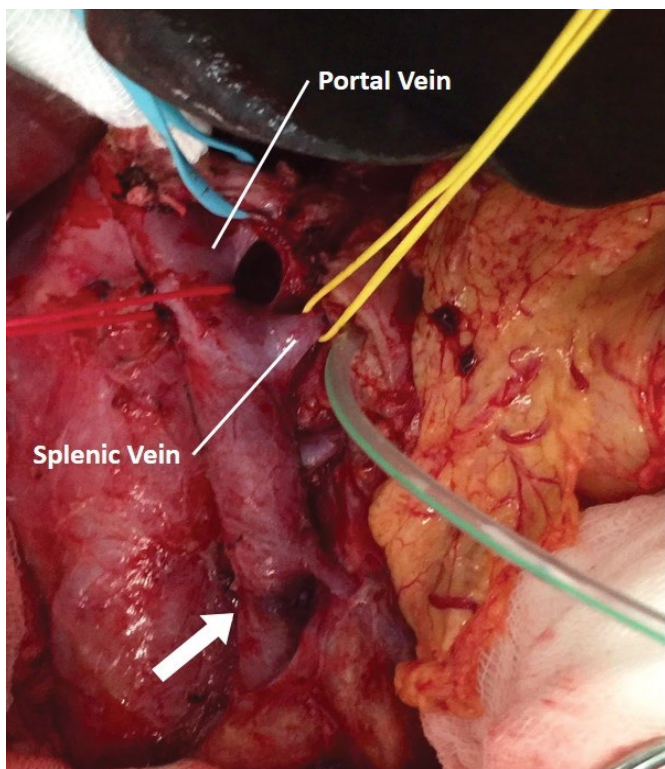
**Figure 1b.** Blood clot inside of the splenic vein (arrow).

Palosaari [2]. This tumor is more prevalent in child population and it is characterized by abdominal pain in most patients, followed by abdominal mass and weight loss [1] with the majority of lesions compromising the head of the gland [4]. Mean age is around 40 years (range 18-80) [8]. There is no gender predominance with this neoplasm and most of the lesions are sporadic. Tumor markers are useful in diagnosis for the pediatric population, but it seems that adult population does not have the same behavior, AFP and CA 19.9 usually are normal [9]. In our first case, AFP levels were elevated, the other case did not have available results. The role for tumor markers in the scenario of adult disease comprises follow-up of those with elevated levels before surgery. Histopathology demonstrates multiple cellular differentiations including acinar, and neuroendocrine lineage, the presence of squamoid corpuscles is the most characteristic feature which varies in different areas of the tumor [6, 10, 11].

Resection is the primary treatment and is feasible depending on the location of the tumor; distal



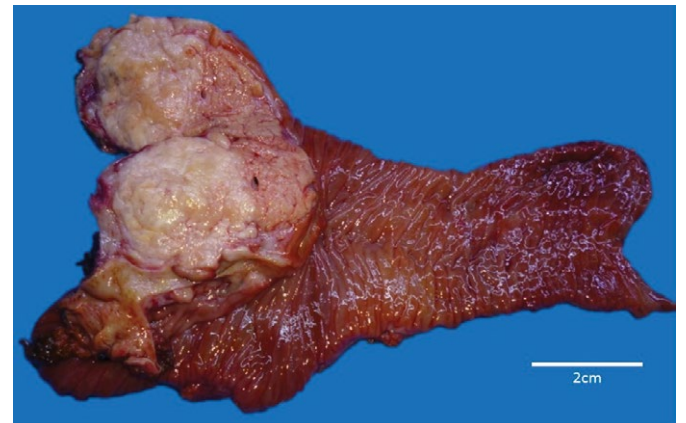
**Figure 2b.** Tumoral thrombus in the mesenteric vein, the specimen was resected *en bloc* with a segment of the vein.



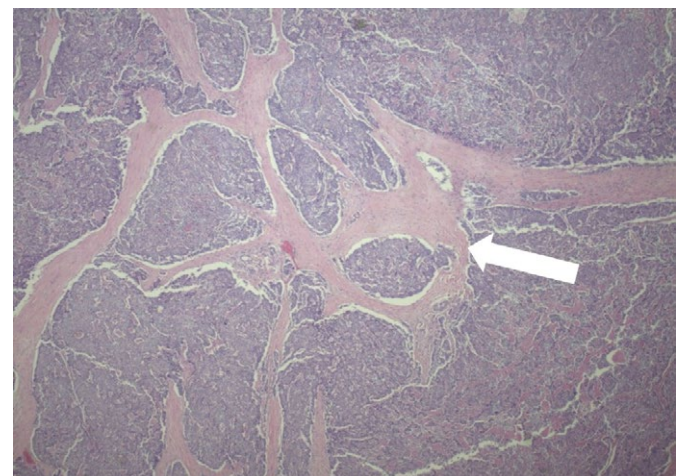
**Figure 2c.** Mesenteric vein reconstruction. Good patency of venous flow was achieved.

pancreatectomy can be done if the tumor is in the body or tail of the pancreas, and pancreaticoduodenectomy can be done for tumors of the head [5]. Complete resection ranges from 66 to 80% [12], but recurrence after resection remains as the main adverse outcome factor. Pancreatoblastoma in adults portends a worse prognosis when compared to child population. The first case had a large tumor and recurrence was within seven months even though an extended resection was performed and negative surgical

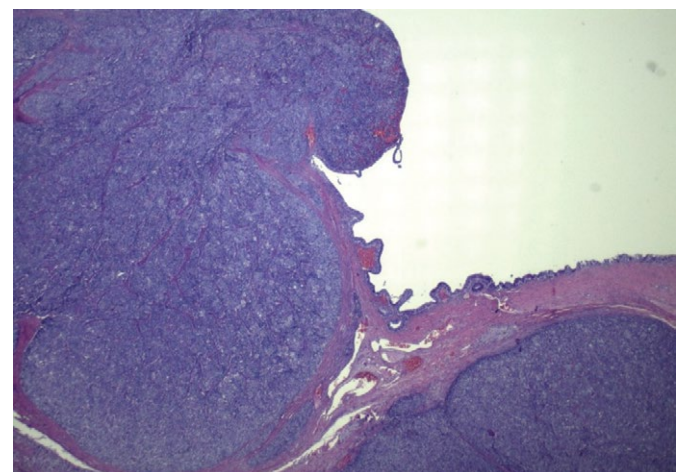
margins were achieved. The second patient is disease-free despite of tumoral thrombus in the portal vein. Since there are different clinical outcomes, surgery remains the only standard treatment. Other approaches might be explored in order to achieve better survival results. This type of tumors, although rare, should be considered in differential diagnosis. The role of chemotherapy is for unresectable, metastatic, or recurrent disease; optimal treatment strategy has yet to be standardized. Treatment



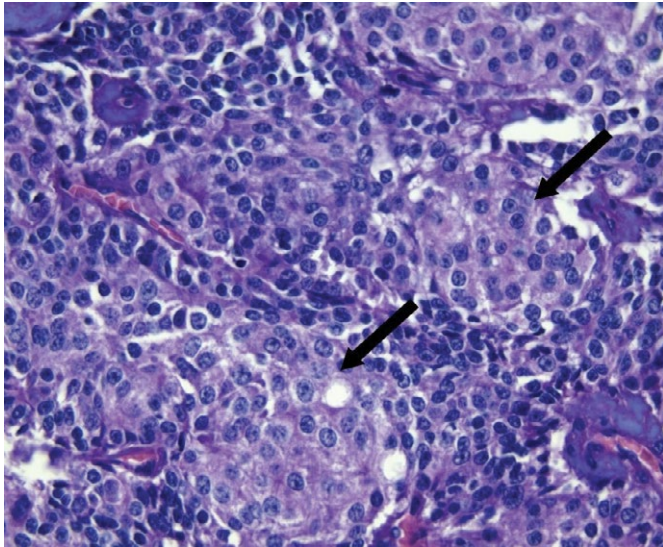
**Figure 3a.** Gross specimen shows a 4 cm tumor mass located in the head of the pancreas, multilobulated surface, partially defined borders. The tumor infiltrated and obstructed the intrapancreatic choledochus.



**Figure 3b.** Nodular and trabecular pattern, fibrous septa dividing them. Neoplasia of nodular and trabecular pattern, separated by fibrous and hyalinized septa (arrow).



**Figure 3c.** Microphotograph of the bile duct infiltration in case 2.



**Figure 3d.** Microphotograph of a squamous nest found in case 2. The immunohistochemistry was positive for EMA, beta catenin and cyclin D1.

options are usually based in children and adolescents reports [5, 13] since there are few cases in adults. Patients initiate gemcitabine, as used in other types of pancreatic cancer and reports of its use in second-line in adults [5], they can receive up to 4 courses. If disease progression is found, 3 courses of cisplatin and doxorubicin are initiated [13]. Palliative oral cyclophosphamide is considered if progression of disease is found after these modalities [14].

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### Conflict of Interest

The authors have declared that no competing interests exist.

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