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## Adult pancreatoblastoma: Current concepts in pathology

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### Abstract

Adult pancreatoblastoma is an exceptionally rare malignant tumour of the pancreas that mimics other solid cellular neoplasms of the pancreas, which may pose diagnostic difficulties. Because of its rarity, little is known about its clinical and pathologic features. This article reviews the clinical and pathologic features of pancreatoblastoma in adults including differential diagnosis, treatment, and follow-up. Although pancreatoblastoma commonly occurs in childhood, there have now been more than 70 adult pancreatoblastomas described in the literature. There is a slight male predominance. There are no symptoms unique to pancreatoblastomas and adult patients are frequently symptomatic. The most common presenting symptom is abdominal pain. Grossly, the tumours are often large and well-circumscribed. Microscopically, pancreatoblastomas are composed of neoplastic cells with predominantly acinar differentiation and characteristic squamoid nests. These tumours are positive for trypsin, chymotrypsin, lipase, and BCL10. Loss of heterozygosity on chromosome 11p is the most common molecular alteration in pancreatoblastomas. Adult pancreatoblastomas are aggressive tumours with frequent local invasion, recurrence, and distant metastasis. Treatment consists of surgical resection. Chemotherapy and radiotherapy may have a role in the treatment of recurrent, residual, unresectable, and metastatic disease. It is important to distinguish pancreatoblastomas from morphological mimics such as acinar cell carcinomas, solid pseudopapillary neoplasms, and pancreatic neuroendocrine neoplasms.

**Key Words:** Pancreas; Adult pancreatoblastoma; Pancreatic cancer; Solid pancreatic mass; Non-ductal pancreatic tumours

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**Core Tip:** Adult pancreatoblastomas are extremely rare tumours of the pancreas. They are composed of neoplastic cells with multiple lines of differentiation and characteristic

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squamoid nests. They mimic other neoplasms of the pancreas, which may give rise to diagnostic difficulties. This article provides an up-to-date review of the clinical and pathologic features of pancreatoblastoma in adults, including differential diagnosis, treatment, and follow-up.

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## INTRODUCTION

Pancreatoblastoma is a malignant epithelial neoplasm of the pancreas composed of cells with predominantly acinar differentiation and characteristic squamoid nests. Neuroendocrine, ductal and less commonly, mesenchymal differentiation can be seen but are often less extensive[1-3]. Less than 1% of pancreatic neoplasms are pancreatoblastomas[4,5]. Pancreatoblastoma commonly occurs in childhood, accounting for 25% of pancreatic neoplasms occurring in the first decade of life, with a mean age of approximately 4 years[1].

Adult pancreatoblastoma is extremely rare. Hence, little is known about its clinical and pathologic features. Furthermore, pre-operative diagnosis can be quite challenging because of the considerable overlap with other cellular neoplasms of the pancreas.

This article provides an up-to-date review of the clinical and pathologic features of pancreatoblastoma in adults, including cytology, molecular pathology, differential diagnosis, treatment, and follow-up.

## EPIDEMIOLOGY

Adult pancreatoblastomas are exceptionally rare. To date, only 74 adult pancreatoblastomas have been reported in the literature, mostly in the form of isolated case reports and small series[6-10]. The mean age at diagnosis is 41 years (range, 18-78 years). There is a slight male predilection, with a male-to-female ratio of 1.2:1.

## AETIOLOGY

The aetiology is unknown. Although most tumours are sporadic[11-15], few adult pancreatoblastomas have been described in the setting of familial adenomatous polyposis (FAP)[8,9]. Rare cases in children have been associated with Beckwith-Wiedemann syndrome[16,17].

## CLINICAL PRESENTATION

Most patients are symptomatic, with very few cases discovered incidentally during routine examination and imaging[4,10,11]. There are no symptoms unique to pancreatoblastomas. The most common presenting symptom is abdominal pain[2]. Other clinical features include abdominal mass, weight loss, nausea, jaundice, and diarrhoea [6,14,15,18,19]. Rarely, patients may present with upper gastrointestinal bleeding[12].

Most adult pancreatoblastomas arise in the head of the pancreas. Of the 74 adult pancreatoblastomas described in the literature, localization data were available in 69 cases. The head of the pancreas was involved in 52.1% of cases (36 patients); the tail in 30.4% of cases (21 patients); the body in 14.5% of cases (10 patients); the body and tail in 1.5% of cases (1 patient); and the ampulla of Vater in 1.5% of cases (1 patient).

Elevated serum levels of CA19-9[14,15] as well as corticotropin releasing hormone secretion[20] rarely occurs in adult pancreatoblastomas. Serum alpha fetoprotein

(AFP) is elevated in some pancreatoblastomas[4,7,13,21,22]. In addition, AFP may be detected immunohistochemically in tumours associated with elevated serum levels of AFP[3,4,22]. Serum AFP is frequently elevated in children[4,10,13] with levels often in excess of 1000 µg/L[3]. In contrast, AFP is not consistently elevated in adults[2,7,10,13]. When present, elevated serum AFP has been used as a marker of tumour recurrence or disease progression because AFP levels should decrease or normalize with successful treatment[7,13,21].

It is important to note that elevated AFP is not specific for pancreatoblastoma in a patient with a pancreatic mass. Pancreatic ductal adenocarcinomas[23] and pancreatic acinar cell carcinomas[24,25] have been associated with elevated serum AFP. Furthermore, AFP is widely used as a tumour marker for hepatocellular carcinoma (HCC). However, the limitations of AFP in detecting HCC includes the poor sensitivity in detecting small tumours and elevated levels of AFP in patients with chronic liver disease without HCC. To overcome this limitation, the *Lens culinaris* agglutinin-reactive AFP (AFP-L3) has been found to be highly specific and useful not only for early detection of HCC but also for predicting the risk of development of HCC in patients with chronic liver disease[26,27]. However, AFP-L3 or other isoforms of AFP are yet to be extensively studied in pancreatoblastomas.

Malignant behaviour is prominent in adult pancreatoblastomas. Approximately 59% of adult patients with pancreatoblastoma develop metastases at the time of diagnosis or afterwards in the course of the disease. The liver is the most common site of metastasis[2,12,15,28] followed by lymph nodes[4,5,29], and lung[4,5,7,15]. Chest wall[5], breast[15], bone[30], and brain metastases[31] are extremely rare. Tumours can invade adjacent structures such as the duodenum, spleen, common bile duct, portal vein, and superior mesenteric vessels[2,4,7,19,29].

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## IMAGING

There are no significant differences in the imaging findings of adult and paediatric patients[2,32]. Pancreatoblastomas are large well-defined heterogenous masses with low to intermediate signal intensity on T1-weighted images and high signal intensity on T2-weighted images. Enhancement is a common feature on contrast-enhanced computed tomography images and may be present on magnetic resonance imaging. Calcifications when present may be rim-like or clustered[19,33]. On ultrasound, pancreatoblastomas are well-demarcated solid masses inseparable from the pancreas with mixed echogenicity[33].

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## CYTOLOGY

Fine needle aspiration specimens are composed of cellular singly dispersed and/or clustered polygonal cells. The cells have round to oval nuclei with fine chromatin pattern, small indistinct nucleoli, and moderate amounts of amphophilic or eosinophilic cytoplasm[3,9]. Squamoid nests or corpuscles are best appreciated in cell block preparations. They are composed of plump epithelioid cells with abundant cytoplasm[7,9].

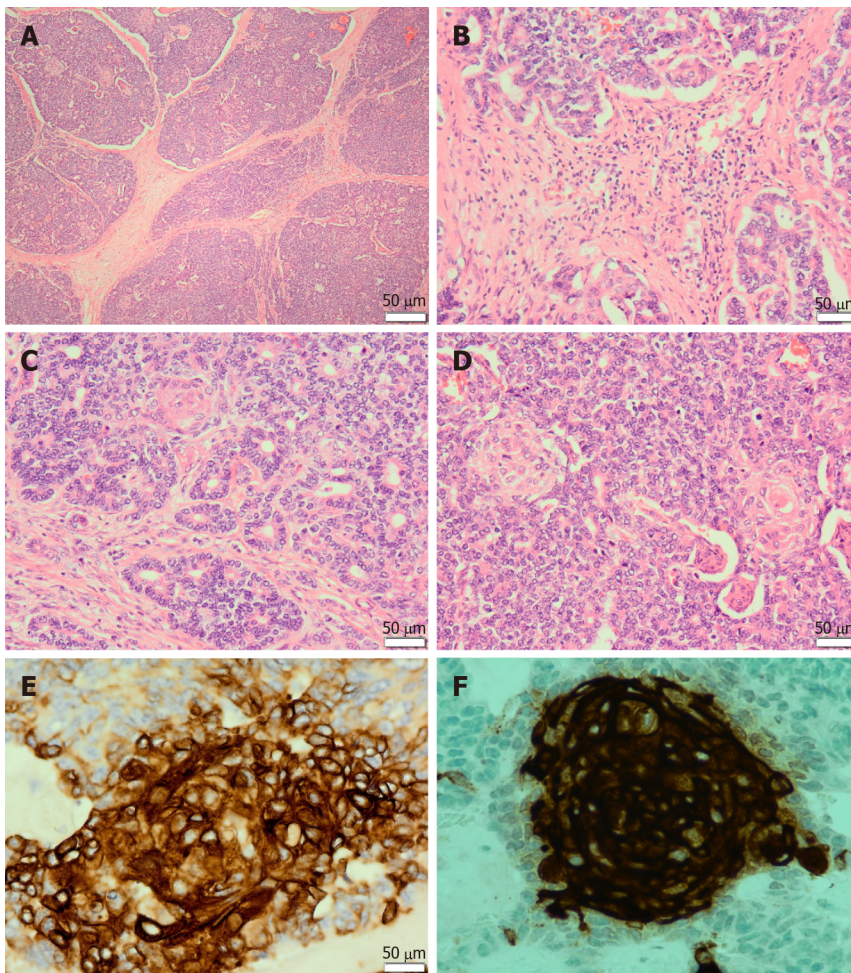
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## PATHOLOGY

Grossly, the tumours are solitary, solid, well-circumscribed, and often encapsulated. Pancreatoblastomas are usually large, averaging 8 cm in diameter (range, 1.8–30 cm)[2,3,6]. On cut section, the tumours have yellow to tan fleshy lobules separated by dense fibrous bands. Foci of haemorrhage and necrosis may be present. Rarely, pancreatoblastomas may undergo cystic change or show gross extension into the adjacent peripancreatic soft tissue[4].

Microscopically, pancreatoblastomas are composed of cellular well-delineated lobules separated by dense fibrous bands, often imparting a geographic low power appearance (Figure 1A). The dense fibrous bands between the lobules are composed of spindled cells with varying amounts of collagen (Figure 1B). Tumours predominantly show acinar differentiation; however, ductal, neuroendocrine and less commonly, mesenchymal differentiation may be present[1-3]. Solid areas with sheets of cells often alternate with areas with acinar differentiation. The acinar units comprise small cells





**Figure 1 Pancreatoblastoma.** A: The tumour is composed of lobules separated by dense fibrous bands, imparting a geographic low power appearance [Haematoxylin and Eosin (H&E) staining, 40 ×]; B: The dense fibrous bands between the lobules are composed of spindled cells with varying amounts of collagen (H&E staining, 200 ×); C: The tumour predominantly shows acinar differentiation. The acinar units are composed of neoplastic cells arranged around central lumina (H&E staining, 200 ×); D: The tumour shows characteristic squamoid nests. Squamoid nests are large islands of plump epithelioid cells with abundant eosinophilic cytoplasm (H&E staining, 200 ×); E: The squamoid nests are immunoreactive for AE1/AE3 (400 ×); F: The tumour shows immunolabeling for CD10 limited to the squamoid nests (400 ×).

with granular cytoplasm arranged around central lumina (Figure 1C). The cells have round to oval nuclei with single prominent nucleoli[1,3,4].

The defining histological feature of pancreatoblastoma is the squamoid nests. Squamoid nests vary from large islands of plump epithelioid cells to whorled nests of spindled cells showing mild to frank keratinization. The cells of the squamoid nests are often distinct from surrounding acinar cells. They are larger than surrounding cells with abundant eosinophilic to clear cytoplasm and without cytological atypia (Figure 1D). The amount of squamoid nests can vary both within and between tumours.

Pancreatoblastomas typically express trypsin, chymotrypsin, lipase, and BCL10. The granules are periodic acid-Schiff (PAS)-positive and resistant to diastase (PASD)[3,4]. Focal expression of chromogranin A and synaptophysin may be present. Squamoid nests may be positive for EMA, AE1/AE3 (Figure 1E) or CD10 (Figure 1F). In addition, patchy nuclear and cytoplasmic expression of  $\beta$ -catenin may be seen.

The staging of pancreatoblastoma follows the TNM classification of carcinoma of the exocrine pancreas[3].

## MOLECULAR PATHOLOGY

Loss of heterozygosity on chromosome 11p is the most common molecular alteration in pancreatoblastomas, occurring in 86% of cases. Molecular alterations in the adenomatous polyposis coli (APC)/ $\beta$ -catenin signalling pathway have also been



identified in 67% of pancreatoblastomas, including biallelic inactivation of the *APC* gene and activating mutations of *CTNNB1* ( $\beta$ -catenin) gene. Biallelic inactivation of the *APC* gene has been identified in a patient with pancreatoblastoma arising in the setting of FAP[8]. Interestingly, aberrations in the APC/ $\beta$ -catenin pathway have been implicated in the development of hepatoblastoma, a tumour associated with Beckwith-Wiedemann syndrome[8,34].

Recent RNA sequencing studies have identified molecular aberrations in the fibroblast growth factor receptor (FGFR) signalling pathway. These include somatic FGFR1 mutation, *FGFR2* gene rearrangement, and a high mRNA expression of fibroblast growth factor (FGF) receptors 1, 3 and 4 as well as of their ligands, FGF3 and FGF4[18].

The most frequent recurrent molecular alterations identified in pancreatic ductal adenocarcinomas, including mutations in *KRAS*, *TP53*, and *CDKN2A/p16* genes, are typically lacking in pancreatoblastomas, suggesting that pancreatoblastomas are genetically distinct from pancreatic ductal adenocarcinomas[8]. Loss of SMAD4/DPC4 expression is rare in pancreatoblastomas[8,35].

## DIFFERENTIAL DIAGNOSIS

Pancreatoblastomas are distinct from the more common pancreatic ductal adenocarcinoma, and it is generally easy to differentiate them on the basis of morphology. The differential diagnosis of pancreatoblastoma includes solid cellular neoplasms of the pancreas such as acinar cell carcinomas, solid pseudopapillary neoplasms, and pancreatic neuroendocrine neoplasms (PanNENs).

There are a number of clinical and morphological similarities between acinar cell carcinomas and pancreatoblastomas. Acinar cell carcinomas are rare, accounting for 1%-2% of pancreatic neoplasms in adults and about 15% in children[1]. Acinar cell carcinomas have a poor prognosis, with a mean survival of 18-24 mo and a 3-year survival rate of 26%[36,37]. Both acinar cell carcinomas and pancreatoblastomas present with non-specific clinical symptoms such as abdominal pain, abdominal mass, and weight loss. In addition, both tumours are cellular neoplasms with acinar differentiation. Neoplastic cells are often polarized around central lumina. The cells contain PASD-positive cytoplasmic granules. Furthermore, acinar cell carcinomas and pancreatoblastomas are typically immunoreactive for trypsin, chymotrypsin, lipase, and BCL10. However, the distinguishing feature is the characteristic squamoid nests seen in pancreatoblastomas.

Solid pseudopapillary neoplasm of the pancreas is a low-grade malignant neoplasm characterized by cells with solid and pseudopapillary growth patterns. Approximately 1%-2% of pancreatic neoplasms are solid pseudopapillary neoplasms, and they frequently occur in girls and young women[3]. Microscopically, solid pseudopapillary neoplasms are composed of poorly cohesive monomorphic epithelial cells arranged around hyalinized fibrovascular stalks, forming solid and pseudopapillary structures. The nuclei frequently show indentations, clefts, and grooves. Typically, these tumours contain scattered PASD-positive hyaline globules, foamy histiocytes, cholesterol clefts, and foreign body giant cells[1]. Solid pseudopapillary neoplasms are positive for nuclear and/or cytoplasmic  $\beta$ -catenin, CD56, CD10, vimentin, and cyclin D1. Unlike pancreatoblastomas, the prognosis of solid pseudopapillary neoplasm of the pancreas is excellent.

PanNENs constitute about 2%-5% of pancreatic neoplasms[3]. They are architecturally diverse and can be confused with pancreatoblastomas. In addition, pancreatoblastomas can focally express neuroendocrine markers. In contrast, pancreatic neuroendocrine tumours are composed of cells with amphophilic to eosinophilic cytoplasm and the nuclei have characteristic salt and pepper chromatin. Typically, pancreatic neuroendocrine tumours strongly express synaptophysin, chromogranin A, and CD56. Features that favour a diagnosis of pancreatoblastoma include predominant acinar differentiation, squamoid nests, PASD-positive cytoplasmic granules, and expression of trypsin, chymotrypsin, lipase, and BCL10.

## OUTCOME

There are no established treatment guidelines for pancreatoblastoma. Treatment consists of surgical resection with a variable combination of chemotherapy, radiotherapy, or targeted therapy (Table 1).

Table 1 Treatment and outcome of adult pancreatoblastoma

Ref.	Treatment	Follow-up (mo)	Outcome
Charlton-Ouw <i>et al</i> [39], 2008	Surgical resection, chemotherapy, RT	60	NED
Levey and Banner[40], 1996	Surgical resection	4	DOD
Palosaari <i>et al</i> [29], 1986	Surgical resection, chemotherapy, RT	15	AWD
Rajpal <i>et al</i> [13], 2006	Surgical resection, chemotherapy	17	DOD
Dunn and Longnecker[41], 1995	Surgical resection, chemotherapy	11	DFUD
Zhu <i>et al</i> [42], 2005	Chemotherapy	9	AWD
Du <i>et al</i> [14], 2003	Surgical resection	6	NED
Hoorens <i>et al</i> [43], 1994	Surgical resection	30	NED
Robin <i>et al</i> [44], 1997	Surgical resection, chemotherapy	7	DOD
Gruppioni <i>et al</i> [45], 2002	Surgical resection	10	NED
Benoist <i>et al</i> [12], 2001	Surgical resection, chemotherapy	36	NED
Mumme <i>et al</i> [46], 2001	Surgical resection, chemotherapy, RT	9	DOD
Salman <i>et al</i> [5], 2013	Surgical resection	30	NED
Salman <i>et al</i> [5], 2013	Surgical resection, chemotherapy, RT	41	NED
Salman <i>et al</i> [5], 2013	Surgical resection, chemotherapy, ablation of liver mets	51	DOD
Hayasaki <i>et al</i> [47], 1999	Surgical resection	15	NED
Sheng <i>et al</i> [48], 2005	Surgical resection, chemotherapy, RT, TACE	26	DOD
Balasundaram <i>et al</i> [15], 2012	Chemotherapy	1	DFUD
Klimstra <i>et al</i> [4], 1995	Surgical resection	5	NED
Klimstra <i>et al</i> [4], 1995	None	5	DOD
Klimstra <i>et al</i> [4], 1995	Surgical resection	10	DOD
Klimstra <i>et al</i> [4], 1995	Surgical resection	15	NED
Klimstra <i>et al</i> [4], 1995	Chemotherapy, RT	38	DOD
Rosebrook <i>et al</i> [32], 2005	Surgical resection	NA	NA
Montemarano <i>et al</i> [19], 2000	Surgical resection	NA	NA
Abraham <i>et al</i> [8], 2001	NA	NA	NA
Abraham <i>et al</i> [8], 2001	NA	NA	NA
Boix <i>et al</i> [20], 2010	Surgical resection	3	DOD
Pitman and Faquin[7], 2004	Surgical resection, chemotherapy, RT	108	AWD
Savastano <i>et al</i> [49], 2009	Surgical resection, chemotherapy, RT	NA	NED
Cavallini <i>et al</i> [10], 2009	Surgical resection	51	NED
Cavallini <i>et al</i> [10], 2009	Surgical resection	15	NED
Hammer and Owens[28], 2013	Surgical resection	NA	NA
Zhang <i>et al</i> [50], 2015	Surgical resection, chemotherapy	NA	NED
Ohike <i>et al</i> [51], 2008	Surgical resection	108	NED
Chen <i>et al</i> [52], 2018	Hepatic transarterial chemoembolization (TACE)	48	DOD
Yamaguchi <i>et al</i> [53], 2018	Surgical resection, chemotherapy	13	DOD
Nunes <i>et al</i> [54], 2018	Palliative care	3	DOD
Vilaverde <i>et al</i> [55], 2016	Surgical resection, chemotherapy	12	DOD
Zouros <i>et al</i> [30], 2015	Surgical resection, chemotherapy, RT	13	DOD
Kuxhaus <i>et al</i> [56], 2005	NA	NA	NA

Comper <i>et al</i> [57], 2009	Surgical resection	NA	NA
Comper <i>et al</i> [57], 2009	Surgical resection	NA	NA
Gringeri <i>et al</i> [58], 2012	Surgical resection, chemotherapy, stereotactic RT	44	NED
Redelman <i>et al</i> [59], 2014	Surgical resection	NA	NA
Tabusso <i>et al</i> [6], 2017	Surgical resection, chemotherapy, RT	10	AWD
Tabusso <i>et al</i> [6], 2017	Surgical resection	15	NED
Liu <i>et al</i> [60], 2020	Surgical resection	24	NED
Reid <i>et al</i> [9], 2019	NA	72.2	DOD
Reid <i>et al</i> [9], 2019	NA	17.9	AWD
Reid <i>et al</i> [9], 2019	NA	3.6	DOD
Reid <i>et al</i> [9], 2019	NA	85	DOD
Reid <i>et al</i> [9], 2019	NA	143.7	DOD
Reid <i>et al</i> [9], 2019	NA	13.6	NED
Reid <i>et al</i> [9], 2019	NA	0.8	DOD
Reid <i>et al</i> [9], 2019	NA	6.5	NED
Reid <i>et al</i> [9], 2019	NA	348	AWD
Reid <i>et al</i> [9], 2019	NA	88	AWD
Reid <i>et al</i> [9], 2019	NA	91	AWD
Terino <i>et al</i> [61], 2018	Chemotherapy	NA	NA
Morrissey <i>et al</i> [62], 2020	Surgical resection, chemotherapy	2	NED
Berger <i>et al</i> [8], 2020	Surgical resection, chemotherapy	18	DOD
Berger <i>et al</i> [8], 2020	Surgical resection, chemotherapy, RT, splenectomy	24	DOD
Berger <i>et al</i> [8], 2020	Surgical resection, chemotherapy, immunotherapy, RT	17	DOD
Berger <i>et al</i> [8], 2020	Chemotherapy, tyrosine kinase inhibitor therapy	15	DOD
Zhang <i>et al</i> [11], 2020	NA	NA	NA
Zhang <i>et al</i> [11], 2020	NA	NA	NA
Zhang <i>et al</i> [11], 2020	NA	NA	NA
Zhang <i>et al</i> [11], 2020	NA	NA	NA
Zhang <i>et al</i> [11], 2020	NA	NA	NA
Zhang <i>et al</i> [11], 2020	NA	NA	NA
Zhang <i>et al</i> [11], 2020	NA	NA	NA
Elghawy <i>et al</i> [31], 2021	Chemotherapy, autologous hematopoietic cell transplantation	57	AWD
Snyder <i>et al</i> [63], 2020	Surgical resection, chemotherapy, GKRS	63	NED

RT: Radiotherapy; GKRS: Gamma knife radiosurgery; DOD: Died of disease; AWD: Alive with disease; DFUD: Died from unrelated disease; NED: No evidence of disease; NA: Not available.

Of the 74 cases of adult pancreatoblastomas described in the literature, outcome data were available in 57 cases. The mean follow-up time was 36 mo (range, 0.8-348 mo). Forty-two percent (24 cases) of patients died of the disease at a mean interval of 27 mo (range, 0.8-143.7 mo); 4% (2 cases) of patients died from unrelated causes (cerebral haemorrhage and pulmonary artery embolus); 16% (9 cases) of patients were alive with disease; and 38% (22 cases) of patients had no evidence of disease (Table 1).

Although long-term survival has been observed in some adults, the prognosis of pancreatoblastoma in children may be more favourable than in adults[1,4,13,14]. Poor prognostic factors include the presence of metastases and unresectable disease[3]. Chemotherapy and radiotherapy may have a role in the treatment of recurrent, residual, unresectable and metastatic disease[3,38]. Because of the tendency for

recurrence and metastasis, long-term follow-up is advised for these patients[38].

## CONCLUSION

In summary, adult pancreatoblastomas are extremely rare. Although these tumours typically occur in children, pancreatoblastomas should be considered in the differential diagnosis of solid pancreatic tumours in adults. An appreciation of distinctive squamoid nests, predominant acinar differentiation, and expression of trypsin, chymotrypsin, lipase, and BCL10 are important for the accurate diagnosis of pancreatoblastomas. These tumours are aggressive with frequent local invasion, recurrence, and distant metastasis. They must be distinguished from morphological mimics. There is a need for further research to better understand the molecular drivers of pancreatoblastomas, identify druggable molecular targets, and, most importantly, improve patient care.

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