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CASE REPORT

Pancreatoblastoma in an Adult Patient. A Case Report

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ABSTRACT

Context Preoperative diagnosis of a pancreatoblastoma in adults is challenging because of its rarity. Furthermore, difficulties increase since pancreatoblastomas share radiological findings similar to those found in other masses of the pancreas. **Case report** A 36-year-old woman was studied with ultrasonography and CT for a mass of the pancreatic head causing obstructive jaundice. Diagnosis of pancreatoblastoma was obtained with histology and immunohistochemistry of the resected specimen. **Conclusion** We reviewed the radiological findings of pancreatoblastomas and possible radiological criteria of differentiation from other pancreatic tumors. A pancreatoblastoma should be considered in the differential diagnosis of a pancreatic mass presenting atypical radiological features.

INTRODUCTION

The term pancreatoblastoma was introduced in 1977 by Horie *et al*. to describe a rare pancreatic tumor, previously known as infantile carcinoma of the pancreas, which shows histological features similar to the pancreatic tissue at approximately the 7th fetal week [1]. A pancreatoblastoma typically affects children, but it can occur throughout one's lifetime; nevertheless, a pancreatoblastoma is extremely rare in adults and, to our best of our knowledge, only 16 cases in adult patients have been reported in the literature [2, 3, 4, 5, 6, 7].

We herein describe an additional case of a pancreatoblastoma in an adult woman, focusing on possible criteria for a differential diagnosis compared with the other more common tumors of the pancreas in non-pediatric patients.

CASE REPORT

A 36-year-old woman was hospitalized for obstructive jaundice. Abdominal ultrasonography showed dilatation of the biliary tree and a mass in the head of the pancreas (Figure 1) measuring 4.3x4.7 cm in diameter and exhibiting a solid but inhomogeneous pattern because of the presence of a hyperechoic component.

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On dynamic computed tomography (CT), the lesion showed well-defined margins and inhomogeneous attenuation due to the presence of both cystic and solid areas, the latter enhancing in the venous phase; dense calcifications were also evident (Figure 2a). The tumor encased the common bile duct; the portal vein was compressed but patent (Figure 2b); there was no radiological evidence of local infiltration, or nodal or distant metastases. Serum levels of tumoral markers (CA 19-9, CA 125, CA 15-3, alpha-fetoprotein and CEA) were within reference limits. A preoperative diagnosis of a solid-pseudopapillary neoplasm of the pancreas was made on the basis of the radiological findings.

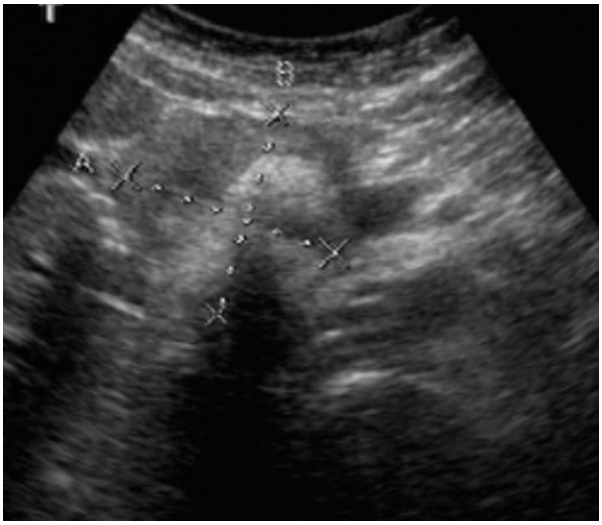

Figure 1. Abdominal ultrasonography reveals a solid mass in the head of the pancreas, with an eccentric hyperechoic component consistent with calcification.

After positioning biliary drainage to treat the jaundice, the patient underwent a laparotomy for a Whipple procedure, but, because the tumor had infiltrated the portal vein, only debulking of the mass was possible. The postoperative course was uneventful.

Grossly, on the cut sections, the tumor was whitish-yellowish, firm and well circumscribed but non-capsulated. At microscopy, the tumor was mainly composed of nests and sheets of small uniform primitive cells frequently showing squamoid nests with central keratinization; occasional ducts were also detected. Several calcifications, mainly arising on the squamoid corpuscles, were also seen (Figure 3ab). The tumor showed vascular and perineural infiltration and focal infiltration of a peripancreatic lymph node.

Immunohistochemical stains showed positivity for cytokeratin 5-6, MNF116 and AE1/3 whereas cytokeratin 7 and 20, usually staining simple and glandular epithelia, were not expressed. In addition, a few endocrine cells could be demonstrated with the neuroendocrine markers synaptophysin and chromogranin A. CD99 was also expressed; CD10 and alpha-fetoprotein were negative.

A final diagnosis of pancreatoblastoma was made on the basis of typical histological and immuno-histochemical features.

After surgery, the patient was treated with radiation therapy and adjuvant chemotherapy; the patient is currently in good condition without radiological evidence of recurrent tumor or metastases.

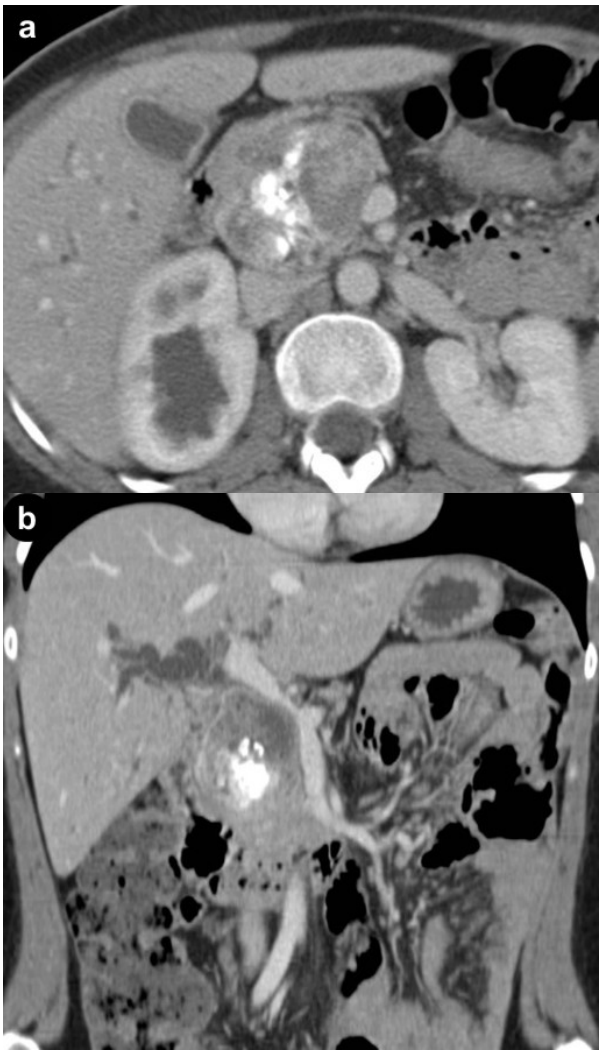

Figure 2. a. On dynamic CT scanning (portal phase), the pancreatic mass is solid, well demarcated and inhomogenously enhanced; dense calcifications are evident; as a collateral finding the right kidney presents pyelectasis from pyelo-ureteral junction syndrome. **b.** Coronal reformation displays an intratumoral cyst-like area superiorly; compression on the portal vein and dilatation of the intrahepatic biliary tree are also visible. Intraoperatively, the tumor infiltrates the portal vein.

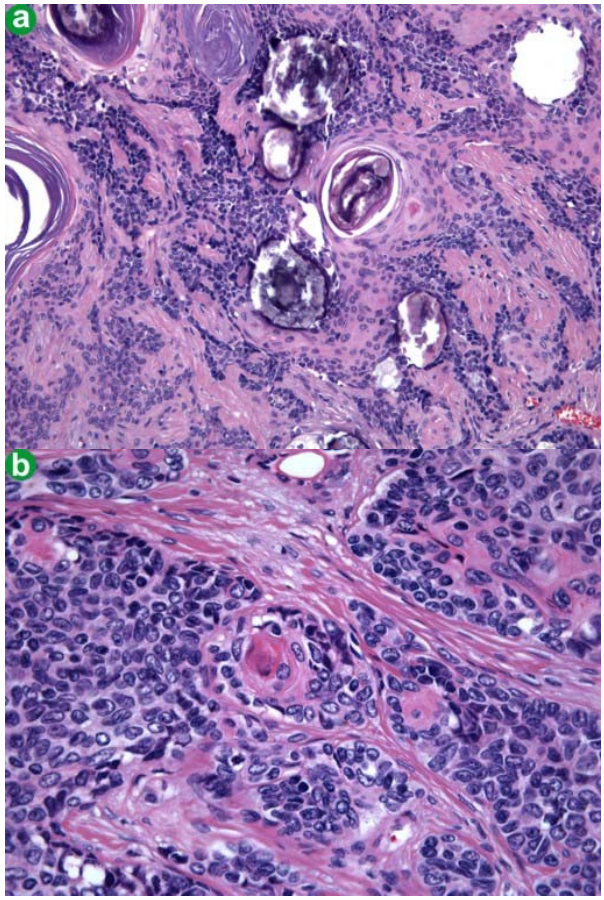

Figure 3. a. Cords of primitive small cells differentiating into squamoid nests with central keratinization and intermixed with several calcifications (hematoxylin-eosin, x400). **b.** Areas with squamoid differentiation without calcification (hematoxylin-eosin, x630).

DISCUSSION

Pancreatoblastoma accounts for 0.5% of exocrine tumors of the pancreas and the highest incidence is found in the first decade of life with a predilection for males and Asians [8, 9]. In approximately two-thirds of cases increased alpha-fetoprotein levels are found, as occur in hepatoblastoma and other blastomal carcinomas; congenital pancreatoblastomas are associated with Beckwith-Wiedemann syndrome [2, 10]. Most pancreatoblastomas exhibit typical genetic alterations similar to those occurring in hepatoblastoma and acinar cell carcinoma, but different from those found in ductal adenocarcinomas [2, 9].

A pancreatoblastoma is a slow growing tumor, soft and well circumscribed, usually very large in size at the time of diagnosis [2, 8, 9]. Approximately one-half of the tumors are found in the pancreatic head [3, 10]. Macroscopically, most pancreatoblastomas are encapsulated and exhibit intratumoral hemorrhage and necrosis; cystic degeneration and calcifications can also be found [10]. The tumor is histologically characterized by different types of epithelial neoplastic cells arranged in lobules separated by gross fibrous stroma; cellular types reflect pathways of differentiation of the embryonal pancreas when the dorsal anlagen and the ventral anlagen fuse [1]. Solid sheets of neoplastic cells with an acinar and squamous-like pattern are evident along with nuclei of keratinization, the so-called “squamoid corpuscles”, which represent the most typical aspect of pancreatoblastoma; immunohistochemistry may show positivity for acinar, ductal and endocrine differentiation [2]. Tumors with overlapping features between pancreatoblastoma and neoplasms with mixed endocrine and solid-pseudopapillary differentiation have rarely been noted [11].

Incidental detection of an abdominal mass is the most frequent clinical presentation of pancreatoblastomas; symptomatic patients usually complain of abdominal pain, weight loss, diarrhea and vomiting; instead, jaundice is rare [3, 4].

Pancreatoblastomas share a similar radiological appearance in both adult and pediatric patients [5]. Most of the tumors are grossly inhomogeneous with all diagnostic imaging modalities due to coexisting solid and cystic areas; septations and calcifications can also be present [3, 6, 8, 12, 13]. In their series of 10 patients, Montemarano *et al*. reported six tumors with well circumscribed margins on CT scans; however, despite the fact that radiological findings may suggest an encapsulated tumor, local infiltration cannot be excluded preoperatively. In pediatric patients, very large pancreatoblastomas can not be differentiated radiologically from hepatoblastoma or other infantile tumors [3].

Solid components and septa are enhanced after the administration of contrast medium on CT and magnetic resonance imaging (MRI). Because of its intrinsic superior capability of tissue characterization, MRI better delineates intratumoral hemorrhage and necrosis areas [3, 5, 6, 8, 12, 13, 14]. On the other hand, CT better demonstrates calcium deposits, which usually appear as rim calcifications or foci of punctuate calcifications [3, 10, 12, 13, 14]. However, as reported in solid-pseudopapillary neoplasms [15], coarse calcifications can be detected. Hepatic, lymph nodal and peritoneal metastases are evident in approximately 30% of cases at clinical presentation; pulmonary and bone metastases have also been reported [2].

Endoscopic ultrasound is useful for the local staging of pancreatic cancer, and for assessing vascular involvement. Although, to our best of our knowledge, there is no report indicating its role in patients with pancreatoblastoma, endoscopic ultrasound is expected to be able to depict the lesion clearly and improve local staging. In addition, endoscopic ultrasound-guided fine needle aspiration, or when required, endoscopic ultrasound-guided tru-cut biopsy, may be performed to clarify the nature of the pancreatic masses [16, 17, 18].

In adults, the radiological differential diagnosis should include pancreatic pseudocyst, mucinous cystic neoplasm, microcystic adenoma, ductal adeno-carcinoma, endocrine neoplasm, acinar cell carcinoma, and solid-pseudopapillary neoplasm; pancreatic and peripancreatic pseudolesions, especially autoimmune pancreatitis and tuberculosis, should also be considered.

A pancreatic pseudocyst can have inhomogeneous contents and peripheral calcification, but a history of pancreatitis and the absence of solid areas leads to the correct diagnosis [3, 19].

Mucinous cystic neoplasms are predominantly cystic; calcifications and fluid-fluid level from hemorrhage can exist but the enhancement of the capsule is typically delayed [19]. Microcystic adenomas, usually found in older women, are characteristically composed of multiple tiny cystic spaces separated by thin septa, eventually calcified and surrounding central stellate scar [3]. Ductal adenocarcinomas are often small in size at the time of diagnosis, and very rarely exhibit necrosis, hemorrhage or calcifications [3]. Acinar cell carcinomas can be very large, well circumscribed and necrotic; metastases are often present, but, unlike pancreatoblastomas, calcifications are not usually evident [2, 20]. Endocrine neoplasms of the pancreas can sometimes be cystic and calcified, but, because they are hypervascularized, they can be diagnosed radiologically by early enhancement on dynamic imaging, even when they are non-functioning [21]. Solid-pseudopapillary neoplasms are low-grade malignancies of the pancreas, usually very large in size, encapsulated, containing solid and cystic areas, and sometimes calcifications; they exhibit a heterogeneous signal on both T1-weighted and T2 weighted MR imaging, and heterogeneous, progressive centripetal enhancement on dynamic imaging [19].

Focal autoimmune pancreatitis can simulate a pancreatic tumor; however, calcifications, vascular involvement and loculation are rare. When localized in the pancreatic head, autoimmune pancreatitis can determine compression and inflammatory changes of the common bile duct [22, 23].

Because of caseation, pancreatic and peripancreatic nodal tuberculosis can show a cystic appearance [22, 24]. It is usually found in immunocompromized patients and in geographic areas of endemic tuberculosis, but nowadays, with globalization, it can be found worldwide.

The prognosis of pancreatoblastoma in the adult population is poor [2]. Surgery remains the treatment of choice for both the primary tumor and the metastases [2, 7]. An empirical approach with chemotherapy can also play a role in the planning of

treatment whereas radiation therapy is indicated in the case of incomplete surgical resection [9].

In conclusion, the preoperative diagnosis of a pancreatoblastoma is very difficult to reach in adults mainly because of its rarity; nevertheless, it should be considered in the differential diagnosis of pancreatic tumors presenting atypical radiological features.

Conflict of interest The authors have no potential conflicts of interest

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