Postpartum Incidental Solid Pseudopapillary Tumor of the Pancreas: Case Report

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ABSTRACT

Solid pseudopapillary tumors of the pancreas are usually described as rare lesions with low malignant potential and unpredictable behavior. The prognosis may be favorable even with local or distant dissemination. Despite the steady increase of cases diagnosed, the pathogenesis of this neoplasm remains unclear. We present a case of an incidental tumor discovered because of an acute abdomen in a postpartum woman without previous evidence of a tumor during the pregnancy.

KEY WORDS: Solid Pseudopapillary Tumor of the Pancreas, Franz Tumor, Cystic Neoplasm of the Pancreas.

INTRODUCTION

In 1959 Franz recognized a rare type of neoplasm of the exocrine pancreas with papillary and cystic characteristics in a 2-year-old boy, who died during an attempted pancreaticoduodenectomy. Since the original description, the frequency of pseudopapillary tumors has been increasing, probably due to the better awareness of these lesions by physicians (1). These tumors account for less than 2% of all pancreatic tumors, and have specific clinical, pathological, and radiographic features that can help to differentiate their diagnosis from other malignant neoplasms of the pancreas (2). These tumors are found predominantly in girls or young women and present as large, encapsulated masses with areas of necrosis, hemorrhage, and cystic degeneration. Until 2002, around 450 cases were reported, most of them in the last 10 years.

CASE REPORT

A 32-year-old woman was admitted to our Institution with a 2-week history of lower abdominal pain, fever, asthenia, and weight loss. She underwent a cesarean delivery 3 weeks before and was treated during 14 days with oral antibiotics due to a supposed endometrial infection. On physical examination, the patient had important right lower quadrant tenderness with peritoneal signs. Laboratory exams revealed significant leukocytosis and the ultrasonography (US) revealed a fluid collection on the right iliac fossa, compatible with acute ruptured appendicitis, and also a 7-cm cyst in the left kidney. She underwent a laparotomy, the diagnosis of acute perforated appendicitis with abscess was confirmed and the treatment performed. An abdominal US on day 5 did not show any complication but, instead of the left kidney cyst as seen in the first US scan, the exam suggested the possibility of a left adrenal neoplasm. However, a computed tomography (CT) scan of the abdomen showed a tumor measuring 7.0-X 6.0-x 6.5-cm in the tail of the pancreas, predominantly cystic, but enhancing a rim of solid tissue, suggesting a mucinous cystadenoma (Figure 1). Carcinoembryonic antigen and Ca-125 tumor markers were within normal limits.

Two months later, she underwent a new laparotomy to remove this tumor. The mass was well-limited and surrounded by large veins, showing close relation to the splenic vein. No signs of distant dissemination or local invasion were seen. The patient underwent a complete surgical resection of the tumor, including a minimal part of the pancreas tail. The spleen was removed in bloc because of vascular damage of the splenic vein (Figure 2). The postoperative course was uneventful and she was discharged from the hospital on day 7. No adjuvant therapy was used and there were no signs of recurrence of the tumor 18 months later.

Macroscopically, the tumor was uniloculated, filled with dark liquid and some clots, suggesting previous bleeding. The inner surface was irregular, with friable trabecula and tanned-gray zones. Light microscopy demonstrated predominantly solid areas with a sheet-like arrangement of polygonal cells and scarce stroma (Figure 3-A), areas with solid and cystic arrangement (Figure 3-B), and areas with predominant pseudopapillary formations (Figure 3-C). No signs of vascular invasion were detected.

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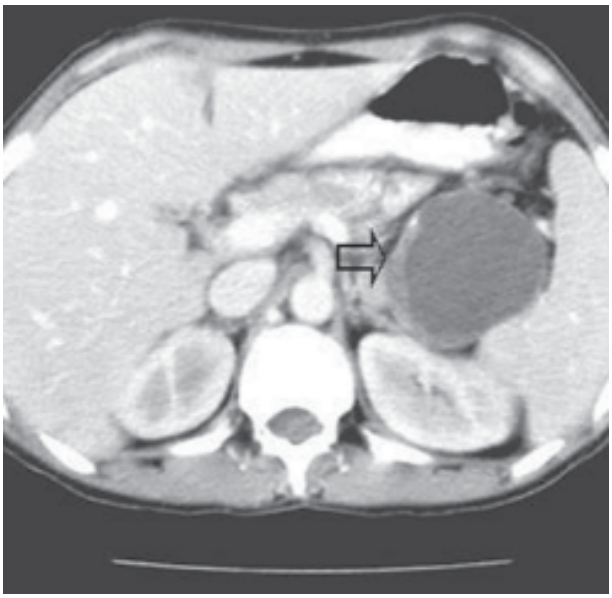


Figure 1 – CT scan of the abdomen shows a 7.0-x 6.5-cm cystic pancreatic mass with an enhancing rim of solid component (arrow).

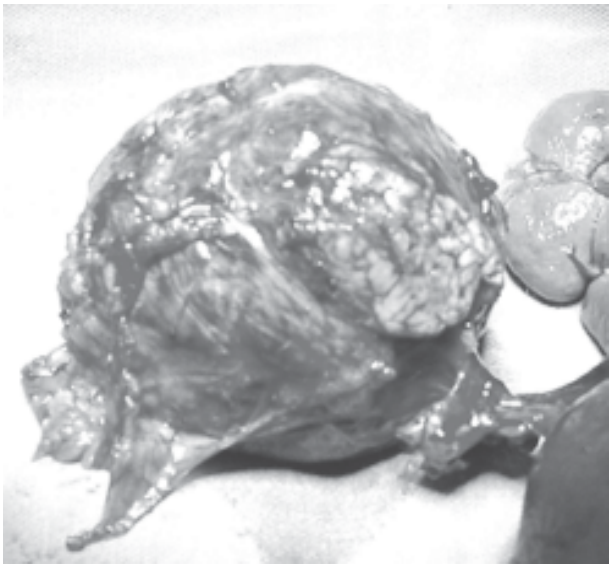


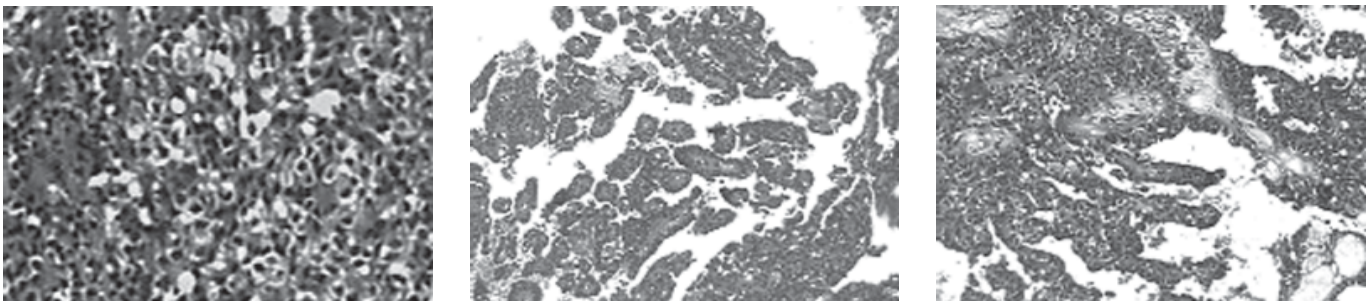
Figure 2 – Macroscopic appearance of resected solid pseudopapillary tumor. The lesion is well limited in close relation to the splenic vein.

DISCUSSION

Before the new classification of pancreatic neoplasm's was created by the World Health Organization in 1996, misdiagnosis between pseudopapillary tumors, adenocarcinoma, islet cell tumors, cystadenomas, papillary cystadenocarcinoma and cystadenocarcinoma were common (3).

There is a clear female predominance (>90%) and most of these tumors are diagnosed in the second and third decades of life. Fifty-two percent of the patients are under 20 at the moment of the diagnosis (4). The distribution of the tumor in the pancreas is the same in all age groups (head and neck, 35%, body, 25%, tail, 40%). The etiopathogeny is unknown, but some authors, based on morphological and immunohistological features, support the idea that they originate from pluripotent embryonic cells, and grow by the action of female hormones. This hypothesis is supported by the presence of progesterone receptors on the epithelia in 90% of the tumors. In contrast, the role of estrogen receptors has shown conflicting results in the presented series (2).

In the present report, there was no clinical evidence of the tumor during the pregnancy, but the US was not directed to this diagnosis. However, sex hormones play an important role in the growth of the solid pseudopapillary tumors and pregnancy is associated with stimulation of tumor growth (6,7).



(A) (B) (C)

Figure 3 – Solid areas with a sheet-like arrangement (A). Areas with solid and cystic components (B). Predominant pseudopapillary formations (C). HE, 100X.

The diagnosis is often casual through an image examination made for other purpose or during the investigation of symptomatic palpable masses in young women, which is responsible for the frequently delayed diagnosis. Mild abdominal pain or early satiety occurs due to compression of neighboring organs. Thus, usually these tumors are quite large at the initial presentation. The median size in a series of 24 patients was 8 cm (range, 1-20). In 8% of the cases, the initial presentation is an acute abdomen (3).

The US usually reveals a well-limited, cystic or solid-cystic mass. The ecogenicity of the tumor is quite variable and depends on the predominance of the cystic (hipoechogenic) or solid (hyperechogenic) component. Depending on the tumor location, preoperative assessment may suggest that it originates from the adrenal glands or that it is a pancreatic endocrine tumor (2).

CT scans of the abdomen usually show a well-encapsulated mass with both solid and cystic components. There may be calcifications in the periphery of the mass and also intravenous contrast enhancement. However, none of these radiographic features is clearly diagnostic. Despite these features, the accuracy of CT scan in characterization of cystic pancreatic masses was about 60% in one series (6). Some authors consider that a correct preoperative diagnosis by CT or US guided aspiration biopsy might be feasible (6). In adults, fine needle biopsies have become a mainstay in the evaluation of pancreatic masses, particularly those that are cystic in nature. The reported sensitivity in the diagnosis of pancreatic malignancies with this technique ranges from 75% to 88% (1). However, some authors consider the biopsies unnecessary due to the risk of extra pancreatic dissemination and because the definitive diagnosis is obtained only on anatomopathological basis (4). Differential diagnosis for cystadenomas, cystadenocarcinomas, islet cell tumors, and pseudocysts is sometimes very difficult. Frozen sections can miss the correct diagnosis, but usually are useful to discard pancreatic adenocarcinoma (1,2).

Around 7-14% of these tumors develop malignant features, with local invasion and distant metastasis (1,2). Since the malignant potential of these tumors is low, prognosis is excellent when complete resection is achieved, even in the presence of metastatic disease. Local recurrences after complete resection are rare. Some authors have defined this tumor as "an infrequent carcinoma of good prognosis". The most common sites of metastatic disease are the liver (42%), lymph nodes (25%) and peritoneal spread (42%). This tumor has a high propensity for peritoneal spread, similar to the ovarian tumors. Fortunately, long–term survival is also possible even with disseminated disease (6). Pathologic factors like mitotic rates; nuclear pleomorphism and vascular invasion were not found to correlate with prognosis. However, an unusual variant with aggressive behavior, high mitotic rates, and increased nuclear pleomorphism was responsible for two deaths in one series (3). Evidence suggests that a translocation between chromosomes 13 and 17 might be a marker of aggressiveness (5). Chemotherapy and hormonal therapy had disappointing results. Isolated reports using high dosages of radiotherapy have shown partial response (3).

The pathologic diagnosis of solid pseudopapillary tumors is based on the presence of characteristic light microscopic features. Solid areas alternating with pseudopapillary formations, evidence of cellular degeneration, including cholesterol clefts and aggregates of foamy histiocytes, nuclear grooves and aggregates of hyaline cytoplasmic globules are found, at least focally (3). Solid pseudopapillary tumors of the pancreas have a characteristic immunophenotype. Markers of acinar (trypsin, chemotrypsin) and ductal (glycoproteins) differentiation are consistently negative. The immunoreactivity to vimentin, á-1-antitrypsin, á-1-antichymotripsin, phospholipase A_2, Leu–M1 and Ki-M1P is usual, but most of the neuroendocrine markers (NSE, synaptophisyn, chromogranin) are negative, whereas others (CD 56) are positive. Therefore, the absence of distinct immunoreactivity pattern lessens any phenotypic relation with the cell lining of the pancreas (2,3).

The excision of the tumor is the treatment of choice and most of these lesions can be managed with local excision or distal pancreatectomy. In the case of tumors in the head of the pancreas, a pancreaticoduodenectomy with pylorus-preserving technique is advised. Liver metastases are resected when possible. Recurrences occur in 2-6% after complete removal (1,2). Even in the reported cases that were considered malignant, the survival of patients was prolonged after adequate surgical resection. Long survival with metastasis or residual disease has been related (4,6).

In summary, solid pseudopapillary pancreatic tumors represent an intermediate group in terms of malignant disease of the pancreas. Although rare, they should always be in mind when a mass in a young woman is assessed. Radical surgical resection is advised to avoid local and distant recurrences.

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