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Pancreatoblastoma in an Older Adult

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Abstract **Case Report**

Introduction Pancreatoblastoma is a malignant embryonal tumor originating from epithelial cells of the exocrine pancreas with frequent local invasion, recurrence, and metastasis. It usually occurs in the first decade of life. ***Clinical Case:*** A 76-year-old female patient, with a 3-month history of a palpable abdominal mass in the left abdomen and weight loss (7 kg in the last month). A CT scan of the abdomen and pelvis reported a tumor in the left kidney measuring 23x11x15cm and a tumor measuring 6x5.6cm in the uterus, which showed a mixed component of probable right ovary origin. Exploratory laparotomy was performed revealing a tumor of approximately 30 cm in diameter with infiltration of the greater omentum, left kidney, left ureter, pancreas tail, and descending colon**.** Histopathological findings were compatible with pancreatoblastoma. ***Conclusion:*** The relevance of considering pancreatoblastomas as a differencial diagnosis in adult patients with retroperitoneal masses needs to be highlighted.

Keywords: Pancreatoblastoma, pancreatic tumors, general surgery, cancer, malignant tumors.

INTRODUCTION

Pancreatoblastoma is a malignant embryonal tumor originating from epithelial cells of the exocrine pancreas with frequent local invasion, recurrence, and metastasis [1, 2]. It occurs in the first decade of life, starting as early as from 5 years [4], it rarely occurs in adults, and has male predominance [3, 4]. It has a size of approximately 2 to 10 cm [5]; with slow growth and is divided according to the presence or lack of epithelium. Furthermore, depending on the epithelial location it is divided into exocrine and endocrine [2].

It is considered a rare tumor according to records of cases currently reported by the Surveillance, Epidemiology and End Results (SEER) program of the National Cancer Institute of the United States, with an incidence of 0.4% [2]. In the PUBMED biomedical database platform, 59 cases were reported in South America between 2000 and 2020. In Ecuador, according to statistics from the Cancer Society, mortality from malignant pancreatic cancer until 2014 corresponds to 3.8% of cases and there is no report of pancreatoblastoma in adults [7].

The differential diagnosis is based on the origin of the epithelium and its location. Tumors of the exocrine epithelium of the pancreas are adenocarcinoma, adenosquamous carcinoma, acinar cell carcinoma, and solid pseudopapillary tumors. Non-epithelial tumors are liposarcomas, teratomas and lymphomas. Endocrine tumors are malignant insulinomas, gastrinomas and glucagonomas [2].

CLINICAL CASE

A 76-year-old female patient, hypertensive under treatment, of low socioeconomic status. He came due to a 3-month history of a palpable abdominal mass in the left abdomen and weight loss (7 kg in the last month). A CT scan of the abdomen and pelvis reported a tumor in the left kidney measuring 23x11x15cm and a tumor measuring 6x5.6cm in the uterus, which showed a mixed component of probable right ovary origin.

Based on this, it was decided to perform an exploratory laparotomy, revealing a tumor of approximately 30 cm in diameter with infiltration of the greater omentum, left kidney, left ureter, pancreas tail, and descending colon. Uterus was found enlarged, the right ovary presented tumoral characteristics, so an abdominal tumor excision + left nephrectomy + left partial hemicolectomy + Hartman-type colostomy + distal pancreatectomy + simple total hysterectomy + bilateral salpingo-oophorectomy was performed.

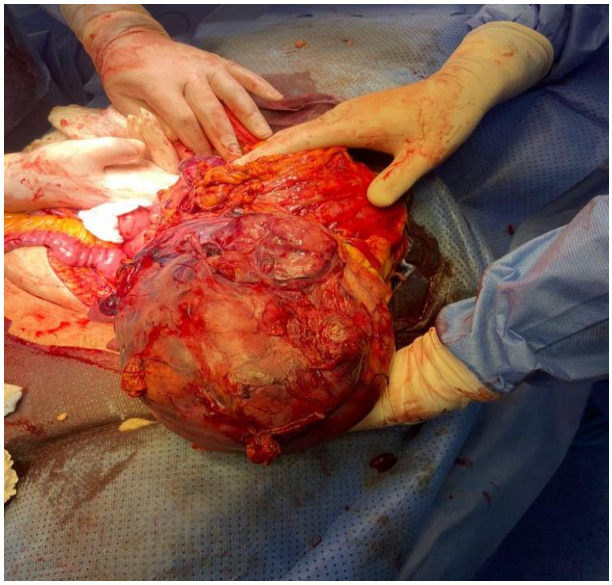

Figure 1: Large multilobulated retroperitoneal tumor with liquid and solid components (predominantly) adhered to large structures

The subsequent histopathological study reported a heterogeneous neoplasm (neuroendocrine component, acinar type, and squamous type nests) with morphology compatible with pancreatoblastoma. Immunohistochemical tests were performed, finding the following as positive: cd 56 - ki67 (in 50% of tumor cells) - chromogranin - synaptophysin - cam 5.2 - broad spectrum keratin - beta catenin - epithelial membrane antigen (in squamous type nests) - cd56 - synaptophysin - cd10 - cytokeratin (ck7) - cam 5.2; and the following as negative: keratin 5/6 - afp alfapheroprotein - inhibin -

Gata 3 - pax 8 - cdx2 - muc4 - thyroid transcription factor - calretinin - progesterone receptors - estrogen receptors - cd99 - cytokeratin (ck20).

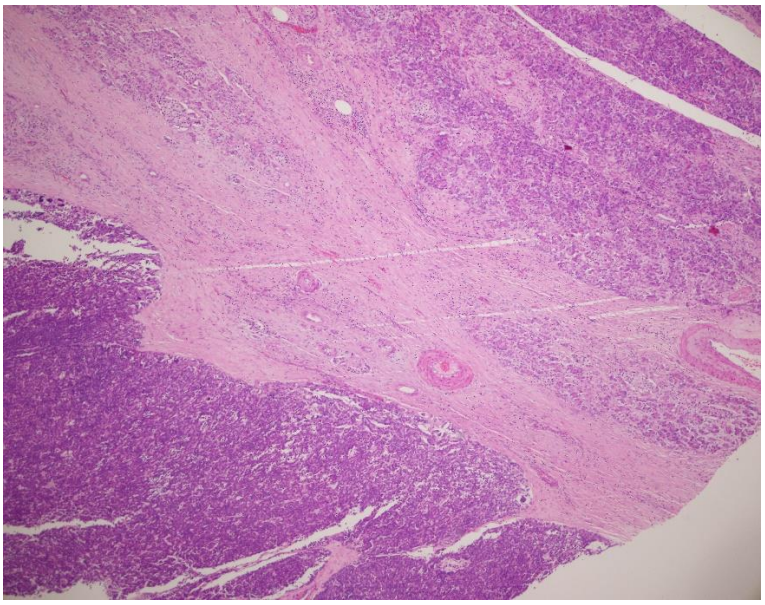

Figure 2: Pancreatoblastoma with its lines of pancreatic differentiation: acinar (predominant), neuroendocrine and squamous type morulae

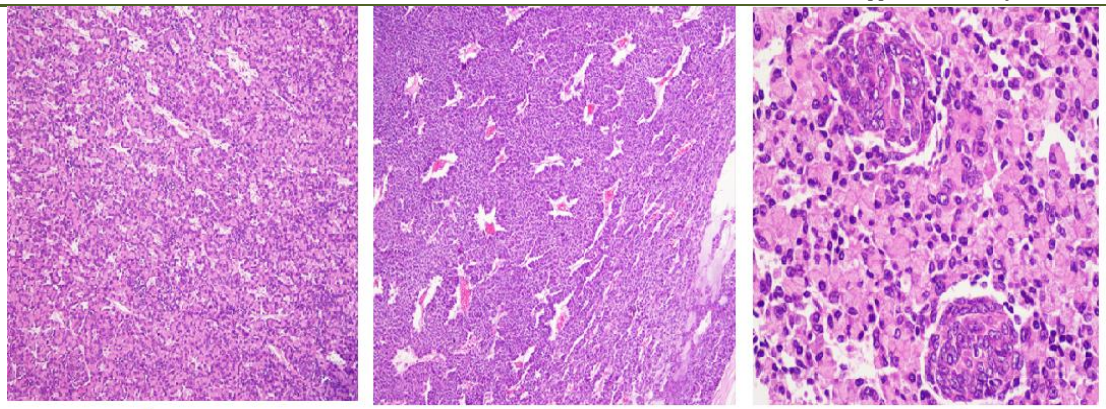

Figure 3: Components of pancreatoblastoma: 1 acinar, 2 neuroendocrine and 3 squamous type morulae

In the postoperative period the patient evolved favorably; she remained hospitalized for 10 days after which was discharged home and initiated the first cycle of chemotherapy.

DISCUSSION

The etiology of pancreatoblastoma is unknown, but it is associated with an embryologic defect of the pancreas; thus, related to hereditary syndromes such as familial adenomatous polyposis (FAP) and Beckwith-Wiedemann [8]; however, this patient did not present any of the aforementioned.

The symptoms are sometimes characterized by abdominal pain [3] and a palpable mass in the epigastrium and mesogastrium, although they are not specific, thefore, resulting generally in a late diagnosis, when the disease is already in an advanced stage [2, 3, 9], as seen in this patient. Jaundice, weight loss, vomiting, diarrhea, or constipation are rarely present [2, 10].

Among imaging studies, the one that stands out the most and is economically profitable is ultrasound, where a mass is usually observed in the head and body of the pancreas, well circumscribed [9], heterogeneous, solid, encapsulated, and cystic [2, 4, 15]. In a CT scan of the abdomen and pelvis, a partially or totally limited pancreatic tumor can be observed with multilobulated borders, cystic areas, and septa with hyperdensity in small parts compatible with calcifications. The most common sites of metastasis are the liver, lymph nodes, lung, brain, in rare cases the omentum, colon, spleen, kidneys and adrenal glands [2, 8, 9]. Also, areas of tumoral invasion may be noticed, as in this case, allowing for a better surgical planning and treatment decision overall.

On the other hand, a primary tumor marker is serum alpha fetus protein, considered in pancreatoblastoma in 75% of cases. When there is liver metastasis or greater severity, elevated lactate dehydrogenase (LDH) is also found in the bloodstream [8]. The patient did not demonstrate the latter, luckily.

The histological diagnosis is by biopsy of the mass with its respective histological examination. Histopathologically, an epithelial tumor with acinar differentiation is observed in this type of tumor [3, 11]. Meanwhile, macroscopically, it appears grayish or brown in color, large, with a soft consistency and cystic changes [3]; in concordance to this case.

Microscopic infiltration of polygonal squamous cells separated by stromal bands, trabeculae and acini is also seen [4, 10, 12, 13]. In this case, the histopathological and immunohistochemical diagnosis reported pancreatoblastoma, ruling out that the tumor was of renal origin and the right adnexal tumor reported serous cystadenoma (benign). Respecting immunohistochemical staining, trypsin staining, cytokeratin AE1/ AE3, CK19, CK7, CK8, Ki-67, CEA can also be seen [6, 8, 12, 14].

Treatment is surgical through complete resection and negative oncological margins. Chemotherapy and radiotherapy are considered in cases of recurrence or metastatic presentation; however, they are not a standardized protocol [2]. In chemotherapy the most used drugs are cyclophosphamide doxorubicin, etoposide and cisplatin [10, 17]. In this case, successful surgical resection of the tumor was performed, but since it was adhered to adjacent structures, their excision had to be performed (tail of the pancreas, descending colon and left kidney), in addition, a simple total hysterectomy was performed for an apparently malignant right adnexal tumor.

The prognosis will depend on whether the tumor is resecable or unresecable and the presence of metastasis. It is good when the tumor is completely removed and poor when there is metastasis. This type of tumor behaves aggressively with an unfavorable response in most cases, in contrast to this patient were it

was found in adulthood; moreover, the patient could survive and receive chemotherapy as a following treatment.

CONCLUSION

The following report presents a rare case of an elderly patient with an abdominal tumor compatible with Pancreatoblastoma, a malignant embryonal tumor originating from epithelial cells of the exocrine pancreas and frequently ocurrying in pediatric patients.

Variable symptoms may be present, being the main a palpable mass in the left abdomen. When this is seen, it has generally already produced biliary obstructive symptoms and is detected in advanced stages.

The diagnosis may be aided by CT, which can show other organ compromises or metastases.

The treatment must be evaluated individually in each patient, always considering a total surgical resection, if the clinical status of the patient and tumoral stage allows for it.

The relevance of considering pancreatoblastomas as a differencial diagnosis in adult patients with retroperitoneal masses needs to be highlighted.

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