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Case Report

Acinar Cell Pancreatic Carcinoma: a Rare Case of Pancreatic Tumor and Short Review of Literature

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Abstract

Pancreatic acinar cell carcinoma (PACC) is a rare subgenre of pancreatic adenocarcinoma, where the cells show acinar architecture. Specified causes of this neoplasia have not yet been deduced and it usually appears with nonspecific symptoms. Diagnosis is based on its characteristics in various imaging techniques, on its histological characteristics and the expression of specific immunohistochemical biomarkers. Surgical excision of the tumor is usually performed with high chances of recurrence, whereas the benefits of radiotherapy and chemotherapy are still ambiguous. In this paper, a 51-year-old female patient with a mass in the head of the pancreas was taken as a case study. She presented with icterus and imaging showed a defined mass in the head of the pancreas accompanied by dilation of the biliary tree. Biopsy and histological assessment done after the surgical excision showed components of PACC differentiation. The patient was regulated after surgery, especially regarding endocrinology and immunology.

Keywords

acinar cell, pancreas, prognosis, rare tumor

INTRODUCTION

Pancreatic cancers can be classified into two major categories, epithelial and nonepithelial. A variation of the former category is pancreatic acinar cell carcinoma (PACC), a type of solid neoplasia characterized by its morphological and functional similarity to normal pancreatic acinar cells.[1-3] PACC is a primarily malignant neoplasia that belongs to the pancreatic adenocarcinoma family; however, documentation of differentiation from the norm is rare.

Despite acinar cells constituting the majority of pancreas' mass, PACCs are disproportionately rare, with about one per cent of pancreatic tumors being of this type. Patients are primarily male, with a male-to-female ratio of 2-3.6:1, about the age of 60, with the median age being 58, whereas race does not seem to play a role. These tumors are usually large, with a mean size of 10 cm at the time of detection, soft, round, and well defined; they have a yellow to brown color and are most frequently located in the head of the pancreas, with a tendency to infiltrate adjacent tissues. Sometimes lesions may even be enclosed in a fibrous capsule, which leads to displacement of near structures.[2,3] Immunohistochemistry is useful to diagnose those kinds of tumors, mainly because of the enzyme-producing cells. Labelling for trypsin, chymotrypsin and lipase are usually positive, whereas amylase is not so often detectable. Other markers include AE1/AE3 and CAM 5.2, which are positive due to CK8 and CK18 production from the cells.[1-3,5,6]

Patients who have this disease usually appear to have non-specific symptoms with weight loss, nausea, abdominal pain, vomiting, cachexia and diarrhea being the most prominent.[1-3] Despite affecting more frequently the head of the pancreas, icterus due to bile obstruction is rare, a key difference with ductal adenocarcinoma of the pancreas.[1,2] There may be evidence of elevated lipase in the blood, but in severe circumstances, a paraneoplastic syndrome known as lipase hypersecretion syndrome – characterized by blood lipase levels above 10,000 U/dl – occurs in roughly 10% to 15% of cases.[1-3,5] Regarding the prognosis of the disease, it is poor as PACC is a malignant and aggressive cancer. Survival is determined mainly by the stage of the cancer and the presence or not of metastases, which most often occur in regional lymph nodes and the liver.[1,2] The 5-year survival is estimated under 10%, but even patients with metastasized cancer commonly show a survival of two-three years.

Like other neoplasias, PACC is characterized by a set of genetic mutations that are found in the neoplastic cells' genome. This type of cancer, in 20-25% of the cases, shares the same pathway mutation as colon cancer, which affects the adenomatous polyposis coli (APC) gene/β-catenin gene.[1-3,5]

Taking into account PACC's special macroscopic, microscopic, immunohistochemical and genetic features, diagnosis is based on laboratory examinations, imaging techniques and biopsy.[1,2] More specifically, the main finding of biochemical examinations is elevated serum lipase values when lipase hypersecretion syndrome is present, whereas biomarker levels are typically normal. Computerized tomography and magnetic resonance imaging are the preferred imaging methods showing demarked, homogenous, and round masses in the pancreas parenchyma, with cystic areas being sometimes present.[1,2] Lastly, biopsy is done using ultrasound guide fine needle aspiration (FNA) method, which usually shows cells with acinar differentiation, often without their cytoplasm which is characterized by high fragility.[1,2]

Regarding treatment, surgical excision is almost always used but with high rates of recurrence of up to 70%, 25% of which are distant metastases. Surgical operation on metastases is proven to ameliorate survival. Benefits of radio and chemotherapy are still debatable and specific treatment schemes have yet to be developed.[1,2]

Herein, we present a case of a female patient with pancreas cancer, which was surgically removed, and later biopsy showed its acinar characteristics.

CASE PRESENTATION

A 51-year-old female patient presented to the hospital due to painless obstructive icterus, accompanied by severe itching. The patient reported some episodes of diffuse abdominal pain accompanied by vomiting the month before she came to the hospital. Biochemical examination results showed elevated alkaline phosphatase (214 U/L) and elevated bilirubin (13.8 mg/dL), with predominance of direct bilirubin (9.4 mg/dL). In a previous hospitalization, ultrasound imaging of the upper abdomen showed a nonhomogeneous hypoechoic mass in the head of the pancreas, mild thickening of the gallbladder's and intrahepatic bile ducts' walls, and a dilated common bile duct, whereas a later MRI-magnetic resonance cholangiopancreatography (MRCP) confirmed the ultrasound's results ***(Fig. 1)****.*

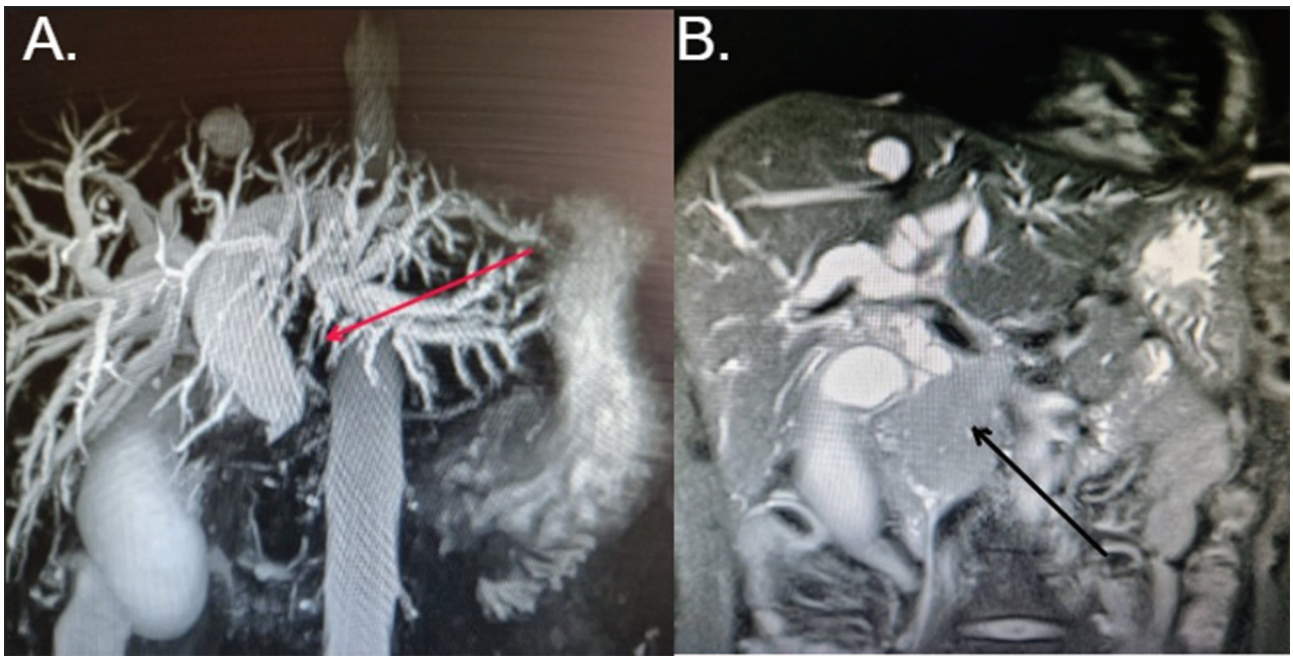

Figure 1. A. Double duct sign with beak-like appearance of the distal bile duct (red arrow); **B**. MRCP revealed tumor at the head of the pancreas with slight enhancement in arterial phase and persistent enhancement in portal phase (black arrow).

During the present hospitalization, a CT-scan of the upper abdomen was performed showing also a protruding mass in the head of the pancreas, and specifically in the uncinate process of the pancreas. The mass seemed to be extending behind the upper mesenteric blood vessels. Dilation of the intrahepatic and extrahepatic biliary tree, with obstruction of the common bile duct was once again found.

Surgical treatment of the patient was decided, and she underwent Whipple surgery along with en bloc pancreatectomy and splenectomy due to the soft texture of the pancreas. A small portion of the stomach was also removed. The removed tissue was sent for macroscopic and microscopic histological examination.

A well-defined grayish neoplasia was found in the head of the pancreas, measuring 4.5×4×3.5 cm and having a lobular appearance. The mass was deemed a low-differentiated adenocarcinoma which showed regional acinar type differentiation and immunohistochemistry, being AE1/AE3+, Cam 5.2+, CK7+, CK19+, α1-ACT+, and regionally BCL-10+. Synaptophysin, S100, and Vim biomarkers were negative. Cells presented with a singular round shaped nucleus with apparent nucleoli and plenty of granular cytoplasm and showed lobular and more rarely tubular architecture. The tumor had a lackluster stroma, and no neuroendocrine component was found. The neoplasia showed a tendency for infiltration of the common bile duct, the mesenteric blood vessels, the peripancreatic fat, and two of the 11 lymph nodes that were removed were found to be infiltrated by cancerous cells. Other than the main mass, the rest of the pancreas parenchyma showed regions of low-grade PanIN and regions of enzymatic necrosis. All of the aforementioned findings indicate that it was a case of PACC, a special type of adenocarcinoma developing from the cells of that produce pancreatic enzymes.

Adding to the surgery, on the fifth postoperative day, an endocrinologic assessment was performed for future regulation due to the excision of the pancreas, which included insulin and pancreatic enzyme substitution and appropriate vaccination due to the splenectomy that was also performed.

DISCUSSION

When considering this particular case several points of interest can be distinguished. The size of the lesion was quite small at 4.5×4×3.5 cm when the median size during diagnosis was about 10 cm, signifying the early diagnosis that was achieved.[2,3] Additionally, the patient presenting with icterus as the main symptom of this clinical entity is interesting, as such a presentation is observed in about 12% of the cases.[2] The architecture that the cells adopted is intriguing, as it combined the common acinar pattern with the more rare tubular pattern of development.[2] Immunohistochemically, the presented case abides by the most common biomarkers, showing AE1/AE3 and Cam 5.2 positivity, but the CK7 and CK19 positivity that is documented is different from the CK8 and CK18 results that are characteristic of PACC.

Despite it being such a rare clinical entity, there is adequate coverage of PACC in the literature, with a variety of cases having been presented. Kimura et al. and Zundler et al. both describe their separate cases of PACC, each with intriguing characteristics.[7,8] Wang et al., over the span of 10 years, compared 19 cases of PACC with 19 cases of ductal adenocarcinoma of the pancreas. These cases presented interesting differences but also important similarities indicative of the presentation of PACC. All the patients were treated, if possible, surgically, and an overall comparison of the two pathologies indicated a better prognosis for DAC.[9]

All things considered, PACC is a rare disease that physicians must take into account when making a differential diagnosis because it is not as obvious how it differs from other pancreatic neoplastic pathologies. This case's unique characteristics highlight the variability of PACC, regarding prevalence, clinical presentation, and immunohistochemistry, which further incommodes a correct diagnosis. Therefore, the presence or absence of the disease, the main parameter that is going to determine the patient's well-being due to PACC's bad prognosis, should be justified by any means necessary.

CONCLUSION

To conclude, pancreatic acinar cell carcinoma can be considered a rare subtype of adenocarcinoma of the pancreas, usually occurring in the head of the pancreas, and presenting itself with nonspecific symptoms. It is also possible for it to appear in other parts of the pancreas as in other organs of the abdomen. In our case, a 51-year-old female patient, presenting with painless icterus, had a mass in the head of the pancreas which was later deduced to be PACC. After the surgery, the patient was regulated regarding her endocrine system and her immune system.

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Competing interests

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Author contributions

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A.S., and D.C.: analysis and interpretation of data; T.T.: drafting the article or reviewing it critically for important intellectual content; T.T.: final approval of the version to be published; D.L. and D.C.: agreement to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

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Ацинарно-клеточная карцинома поджелудочной железы: редкий случай опухоли поджелудочной железы и краткий обзор литературы

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Резюме

Ацинарно-клеточная карцинома поджелудочной железы (АККПЖ) – редкий подтип аденокарциномы поджелудочной железы, клетки которой имеют ацинарную архитектуру. Конкретные причины этой неоплазии ещё не установлены, и она обычно проявляется неспецифическими симптомами. Диагноз основывается на его характеристиках при различных методах визуализации, гистологических характеристиках и экспрессии специфических иммуногистохимических биомаркеров. Хирургическое иссечение опухоли обычно проводится при высокой вероятности рецидива, в то время как преимущества лучевой и химиотерапии до сих пор остаются спорными. В этой статье мы представляем пациентку 51 года с образованием головки поджелудочной железы. У неё была желтуха, и визуализация показала определённое образование в головке поджелудочной железы, сопровождающееся расширением жёлчного дерева. Биопсия и гистологическая оценка, выполненные после хирургического иссечения, выявили дифференцировку АККПЖ. В послеоперационном периоде состояние пациента стабилизировалось, особенно с точки зрения эндокринологии и иммунологии.

Ключевые слова

ацинарная клетка, поджелудочная железа, прогноз, редкая опухоль