Primary adenosquamous cell carcinoma of the pancreas: the use of endoscopic ultrasound guided – fine needle aspiration to establish a definitive cytologic diagnosis

Carcinoma adenoescamoso primario de páncreas: uso de la técnica de biopsia por aspiración con aguja fina guiada por ultrasonido endoscópico para establecer un diagnóstico citológico definitivo

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ABSTRACT
Pancreatic cancer is the second most common malignancy of the gastrointestinal tract in the US, and adenocarcinoma has been identified as the most common type of pancreatic cancer. Different types of pancreatic cancers have been classified: adenocarcinoma, ductal adenosquamous carcinoma, solid pseudopapillary tumors, endocrine neoplasms, acinar cell carcinoma, squamous cell carcinoma, cystic tumors, primary lymphoma of the pancreas, and metastatic lesions of the pancreas. Adenosquamous carcinoma is extremely rare, behave in a very aggressive way and is responsible for the 1 to 4% of the pancreatic exocrine neoplastic lesions. We describe the case of an 82-years-old African American female, presenting to our institution with quantifiable weight loss (12 kg in 3 months), jaundice and abdominal pain. On admission, laboratory tests were obtained: total bilirubin: 11.07 mg/dl with a direct fraction of 10.32 mg/dl. Cross-sectional abdominal CT scan with contrast, showed a lesion localized in the pancreatic head (hypodense on T1, measuring 3.5 x 3.5 x 2.5 cm), with vascular invasion of the portal vein. EUS showed a solid, hypoechoic, not well-defined lesion (measuring 3.98 x 3.80 cm), localized between the head and neck of the pancreas. EUS-FNA was performed with a 22G needle using the fanning technique. The cytological specimens demonstrated components of both squamous carcinoma and adenocarcinoma. The patient underwent ERCP procedure, and biliary drainage was performed with an entirely covered metallic stent placement. After a month from the procedures, the patient died due to the severity of the disease. Endoscopic ultrasound has proven to be the best method to diagnose solid pancreatic lesions, including rare and aggressive type of tumors like primary adenosquamous cell carcinoma that we described in this very interesting case report.

Keywords: Endoscopic Ultrasound-Guided Fine Needle Aspiration; Carcinoma, squamous cell; Pancreatic neoplasms (source: MeSH NLM).

RESUMEN
El cáncer de páncreas es la segunda neoplasia maligna más común del tracto gastrointestinal en los EE.UU. y el adenocarcinoma ha sido identificado como el tipo más común de cáncer de páncreas. Se han clasificado diferentes tipos de cáncer de páncreas: adenocarcinoma, carcinoma adenoescamoso ductal, tumores pseudopapilares sólidos, neoplasias endocrinas, carcinoma de células acinares, carcinoma de células escamosas, tumores quísticos, linfoma primario del páncreas y lesiones metastásicas del páncreas. El carcinoma adenoescamoso es extremadamente raro y se comporta de manera muy agresiva, es responsable del 1 al 4% de las lesiones pancreaticas exocrinas neoplásicas. Presentamos el caso de una mujer afroamericana de 82 años de edad, que fue admitida a nuestra institución con pérdida de peso cuantificable (12 kg en 3 meses), ictericia y dolor abdominal. Al momento de la admisión, se obtuvieron pruebas de laboratorio: bilirrubina total: 11,07 mg/dl con una fracción directa de 10,32 mg/dl. La tomografía computarizada abdominal transversal con contraste mostró una lesión localizada en la cabeza pancreática (hipodensidad en T1, 5 x 3.5 x 2.5 cm) con invasión vascular de la vena porta. El ultrasonido endoscópico mostró una lesión sólida, hipoeicoica, no bien definida de 3,98 x 3,80 cm, localizada entre la cabeza y el cuello del páncreas. La biopsia por aspiración con aguja fina guiada por ultrasonido endoscópico se realizó con una aguja 22G utilizando la técnica de ventilación (Fanning). Los especímenes citológicos demostraron componentes de carcinoma escamoso y adenocarcinoma. El paciente se sometió a CPRE y el drenaje biliar se realizó con una prótesis metálica completamente cubierta. Después de un mes de los procedimientos, el paciente falleció debido a la gravedad de la enfermedad. El ultrasonido endoscópico ha demostrado ser el mejor método para diagnosticar lesiones pancreaticas sólidas, incluyendo tumores raros y agresivos como el carcinoma primario de células adenoescamosas que descriptimos en este interesante relato de caso.

Palabras clave: Biopsia por aspiración con aguja fina guiada por ultrasonido endoscópico; Carcinoma de células escamosas; Cáncer de páncreas (fuente: DeCS BIREME).
INTRODUCTION

Pancreatic cancer is the second most common malignancy of the gastrointestinal tract in the United States, and the adenocarcinoma has been identified as the most common type of pancreatic cancer (1). Different types of pancreatic neoplasias have been classified: adenocarcinoma, ductal adenosquamous carcinoma, solid pseudopapillary tumors, endocrine neoplasms, acinar cell carcinoma, squamous cell carcinoma, cystic tumors, primary lymphoma of the pancreas, and metastatic lesions of the pancreas (2).

Ductal adenocarcinoma is present in about 80-85% of the pancreatic tumors cases (3,4). Adenosquamous carcinoma is extremely rare, behave in a very aggressive way and is responsible for the 1 to 4% of the pancreatic exocrine neoplastic lesions (5,6).

This case report illustrates an exotic and rare clinical scenario of a primary adenosquamous carcinoma of the head of the pancreas, presenting with jaundice from distal biliary duct compression.

CASE REPORT

This is the case of an 82-years-old African American female, presenting to our university institution emergency room with quantifiable weight loss (12 kg in 3 months), jaundice, fecal acholia, dark urine and abdominal pain. On admission, laboratory tests were obtained: a complete blood count was within normal limits, alkaline phosphatase: 1,233 IU/L, GGT: 243 IU/L, AST: 2,521 U/L, ALT: 246 IU/L, total bilirubin: 11.07 mg/dl with a direct fraction of 10.32 mg/dl. Cross-sectional abdominal CT scan with contrast, showed a lesion localized in the pancreatic head (hypodense on T1, measuring 3.5 x 3.5 x 2.5 cm), pancreatic duct dilatation and a thinner section of the distal bile duct suspicious for compression, causing intra and extra-biliary duct dilatation of 17 mm. The CT scan also showed vascular invasion of the portal vein and the left renal vein, with the presence of mesenteric lymph nodes measuring up to 1.1 cm.

After a multidisciplinary meeting, the patient was scheduled for an EUS-FNA to establish a histopathological diagnosis and ERCP for biliary drainage.

EUS showed a solid, hypoechoic, well-defined lesion with irregular contours (measuring 3.98 x 3.80 cm), localized between the head and neck of the pancreas (Figure 1). EUS also showed main pancreatic duct dilatation (Figure 2).

Invasion of the portal vein was confirmed during the exam. EUS-FNA was performed with a 22G needle using the fanning technique.

The cytological specimens demonstrate components of both, squamous carcinoma and adenocarcinoma. The presence of malignant keratinized squamous cells is the most reliable evidence of squamous differentiation. (Figures 3 and 4).

After this, the patient underwent ERCP for biliary drainage from the neoplastic pancreatic head obstruction, distal segmental hepatic duct thinning was observed. The biliary drainage was performed with an entirely covered metallic stent. The procedure was performed without any complications.

After a week from the biliary drainage procedure, bilirubin levels started to decrease, with patient clinical improvement.

After a month from the procedures, the patient died due to the severity of the disease.
DISCUSSION

Adenocarcinoma is the most common type of pancreatic cancer. Among the different pancreatic tumors types, we have solid or cystic lesions, as well as endocrine and exocrine gland involvement \(^2\). Ductal adenocarcinoma is the cause of about 80-85% of pancreatic tumors \(^3,4\).

Adenosquamous carcinoma is extremely rare, behave in a very aggressive way \(^5\), and is responsible for about the 1 to 4% of the exocrine pancreatic neoplastic lesions \(^5\).

A review of the literature evaluated 6,668 exocrine pancreatic tumors between the years of 1950 – 1985, and only 68 cases (0.01%) were found to be primary adenosquamous cell carcinoma of the pancreas \(^7\).

Interestingly, our case in difference with others we observe that adenosquamous carcinoma affects predominately white man over 60 years old \(^5,8\). The most common presenting symptomatology is abdominal pain, weight loss, and jaundice \(^9\). The most common affected anatomic site is the pancreatic head.

There are no radiologic findings that distinguish these types of tumors from adenocarcinoma \(^5\).
Adenosquamous cell carcinoma histogenesis remains uncertain, but there are several hypotheses, among these the main ones are: a) pre-existing adenocarcinoma transformation to adenosquamous carcinoma by cellular transformation; b) squamous ectopic epithelial that transform to malignant; and c) a stem cell capable of differentiating into either squamous or glandular cell undergoing a malignant change. There is a very small number of patients that undergo surgery, because the disease process is very aggressive and by the time of diagnosis, the tumor is already staged as grade IV.

Surgery does not always benefit the patients with adenosquamous cell carcinoma of the pancreas, mostly for those presenting with any other additional comorbidities and older age.

The prognosis for these patients presenting with adenosquamous cell carcinoma of the pancreas is less favorable in comparison to those presenting with ductal cell carcinoma of the pancreas.

Smit et al. reported that the mean survival revolves around 5 to 7 months in 72 patients with the disease, only five patients had longer mean survival of one year. More literature review over the aggressively of this type of tumor-like Kardon et al., explains that the mean survival is 12.5 months for patients treated with surgical resection and palliative adjuvant chemotherapy, and only three months for patients treated only with palliative chemotherapy.

Our case report emphasizes the literature evidence, demonstrating that EUS-FNA is the gold standard method for histopathological diagnostic of solid pancreatic lesions and that primary adenosquamous cell carcinoma is a very rare disease process that presents in a more aggressive way in comparison to adenocarcinoma.

In conclusion, the endoscopic ultrasound has been proven to be the best method to diagnose solid pancreatic lesions, including the rare and aggressive type of tumors like primary adenosquamous cell carcinoma that we described in this case report.

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