Patients with McCune–Albright syndrome are predisposed to pancreatic cancer

Ricardo Correa, Mary Esquivel

ABSTRACT
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INTRODUCTION

McCune-Albright syndrome (MAS) is a rare disorder defined by the classical association of polycystic fibrous dysplasia, precocious puberty and café au lait spots [1]. McCune-Albright syndrome are also associated with an increased risk of various endocrine and non-endocrine neoplasms, such as thyroid adenoma, GH-secreting pituitary adenomas, adrenal Cushing’s and hepatobiliary and pancreatic tumors. McCune-Albright syndrome is caused by and autosomal dominant activating mutations of the G-protein alpha subunit gene (GNAS) [2].

CASE REPORT

A 55-year-old male with McCune-Albright syndrome with multiple manifestations including polyostotic fibrous dysplasia, thyroid involvement, history of Leydig cell tumor, and cafe-au-lait spot presented with recurrent episodes of pancreatitis with a pancreatic cyst.

The patient’s history is significant for recurrent episodes of pancreatitis. The first one dates back to 1998 and a subsequent episode was documented in 2003. He recalls undergoing upper endoscopy (probably EUS or an ERCP), that demonstrated pancreatic cysts. In this context, the patient denies a history of alcohol consumption or cholelithiasis. During the admission, liver function test and lipase and amylase were within normal limits. Magnetic resonance imaging (MRI) scan of abdomen plus magnetic resonance cholangiopancreatography (MRCP) showed dilatation of the extrahepatic common bile duct and pancreatic duct, with replacement of pancreatic parenchyma by dilated side branches or pancreatic cyst. These findings are concerning for intraductal papillary mucinous neoplasm (mixed type). An upper endoscopy with ultrasound of the pancreatic area was performed and multiple cysts were seen and biopsied. As per pathology report, there was mucin and CEA was 124, no malignancy identified on the cytology.

DISCUSSION

Somatic activating G-protein alpha subunit gene mutations have been reported in various hepatobiliary and pancreatic neoplasm such as hepatocellular adenoma, hepatocellular carcinoma and pancreatic intraductal papillary mucinous neoplasm (IPMN). cAMP pathway is involved in the pathophysiology of this neoplasm. The prevalence of IPMN is poorly known but has been estimated to be only 25 per 100,000 [3]. The IPMN have been described in patient with Carney complex due to inactivating mutations of PKAR1A, one of the main regulators of the cAMP signaling pathway. Idiopathic pancreatitis has been described in patients with MAS [1, 4] and could possibly been explained by IPMN, since mild pancreatitis is a classic mode of discover these neoplasms. In our patient, the pancreatic cyst was found because he had 2 episodes of pancreatitis and imaging was performed. In 2013, small observational study was published where they describe the new association between MAS and pancreatic neoplasm (IPMN) and liver adenomas and choledochal cyst. In that study, 32% of the patients were found to have hepatic, pancreatic or biliary lesions. Three of six patients in the series had numerous branch-duct IPMN. That study strongly suggests that cAMP pathway is involved in IPMN tumorgenesis. Given the long-term malignant potential of IPMN, all MAS patients might be offered routine screening by MRI scan. All detected lesions, should benefit from a multidisciplinary counseling (surgeon and gastrointestinologist) and management with follow, biopsy and surgical indications should be advised. If no lesion is found, MRI scan might be performed every five years [5]. In our patient, biopsy...
failed to demonstrate IPMN. We will follow-up him with imaging.

How will this affect clinical practice?

The patient presented with a pancreatic cyst and recurrent episodes of pancreatitis. This is a red flag due to the association between MAS and IPMN (presented). In this case, patient with evidence of pancreatic cyst since 2004 with a negative biopsy at that time. Screening is recommended every five years in patient without lesions. There is no recommendation on how frequent we should screen patient with lesions that are negative for malignancy. On this patient, it took eight years for re-screening. The new MRI scan showed a suggestive IPMN lesion that was not demonstrated with the biopsy. Continuous screening should be done, probably every two years. With this new information, MRI scan of abdomen in MAS patients with pancreatic/liver alteration should be done every 2–3 years because of the high risk of developing IPMN or hepatobiliary neoplasm.

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Keywords: Endocrine, McCune-Albright syndrome, Pancreatic intraductal papillary mucinous neoplasm

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Ricardo Correa – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Mary Esquivel – Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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