Metastatic Adenocarcinoma Arising from Fibrocalcific Pancreatic Diabetes

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Fibrocalcific pancreatic diabetes (FCPD) is a rare form of ketosis-resistant diabetes in the young (15 to 40 years old) of unknown etiology. It has been observed in tropical and subtropical countries with highest incidence in south India, and is believed to have some association with tropical chronic pancreatitis, malnutrition, toxin exposure (e.g., cassava) and SPINK1 mutation. It is associated with a hundredfold increased risk of pancreatic cancer compared to the general population.1-3

A 43-year-old diabetic male presented with recurrent abdominal pain since 24 years of age, anorexia and weight loss of 8 kg in the last 6 months. Computerized tomography (CT) revealed multiple calculi in the pancreatic head, body and tail; a 38 mm x 38 mm x 32 mm mass in the pancreatic tail; and multiple target lesions in the right lobe of liver (Figure 1). He was diagnosed of

Figure 1. CT scan of the abdomen showing atrophic pancreas, dilated main and accessory pancreatic duct, multiple pancreatic calculi (thick white arrow). There was a mass in the tail of pancreas (thin arrow) and multiple target lesions in the liver (open arrows), suggestive of metastasis.
diabetes at 28 years age, which was well controlled with glimepiride and metformin. In the last 2 years, the patient was shifted to premixed insulin and subsequently multiple subcutaneous insulin injections due to worsening glycemic control, and was on 58 units insulin/kg/day. His HbA1c at the time of admission was 9.6%. Serum CA19-9 was elevated (828.8 U/L, normal value <40 U/L). Tru-cut biopsy of the right lobe of liver revealed moderately differentiated metastatic adenocarcinoma (Figure 2).

CA19-9 is elevated in 70 to 80% of patients with pancreatic cancer. It is predictive of distant metastasis or disease recurrence. Consensus statements have recommended screening for pancreatic cancer in any individual with an increased risk (more than 10-fold), which includes individuals with at least 3 first-degree relatives with pancreatic cancer, Peutz-Jeghers syndrome, familial atypical multiple mole melanoma and hereditary pancreatitis. The association between FCPD and carcinoma of the pancreas is not well-known, with some reports suggesting that patients with FCPD may be at higher risk. This warrants further studies to evaluate cancer risk, and to develop screening tools for early detection of malignancy.

References