Naked Islets of Langerhans are Frequently Found in Patients with Pancreatic Endocrine Neoplasms

R Wilcox, A Noffsinger.
Department of Pathology; University of Chicago Medical Center, Chicago, Illinois.

Background
90% of pancreatic endocrine cells reside within islets of Langerhans. The remainder lie scattered among the acini and along the larger pancreatic ducts. Rarely, isolated, naked islets have been reported outside the pancreas proper, in the peripancreatic fat. In our experience, this phenomenon usually occurs in the distal pancreas, and is more common in patients with pancreatic endocrine neoplasms. Based on this observation, we examined distal pancreatectomy specimens from patients with endocrine neoplasms and other pancreatic pathologies to determine the frequency with which naked islets occur.

Design
29 distal pancreatectomy specimens were reviewed, 14 of which were performed for endocrine tumors, and 15 for other reasons (ductal adenocarcinoma, pancreatic pseudocyst, solid pseudopapillary tumor, mucinous cystadenoma). Naked islets, defined as islets sitting alone in the peripancreatic fat without accompanying ductal or acinar structures, were recorded as present or absent, and their distance from the closest adjacent pancreatic tissue was determined. These foci were clearly delineated from changes of chronic pancreatitis in that they lacked fibrosis or evidence of residual pancreatic structures. The clinical histories from all patients were reviewed. Stains for neuroendocrine markers and specific pancreatic endocrine products were performed in a subset of cases from patients with pancreatic endocrine tumors.

Results
6 of the 14 distal pancreatectomies performed for endocrine tumors showed naked islets (43%). In 2 cases, the islets were present within 1 mm of the pancreatic parenchyma. In the remaining four cases, naked islets extended from 2 mm to over 1 cm beyond the pancreas proper. Immunohistochemical staining showed the same distribution of pancreatic endocrine products with naked islets as in the normal intrapancreatic islets. None of the patients with naked islets had any clinical features suggesting multiple endocrine neoplasia syndrome, nor were any of the tumors clinically functional. However, the 2 patients with the most extensive distribution of naked islets were the only 2 patients who were symptomatic (nausea, vomiting, night sweats). Among the 15 pancreata resected for non-endocrine lesions, 2 showed small foci of naked islets within 1 mm of the pancreas proper.

Conclusions
• Naked islets are seen in a small proportion of distal pancreatectomy specimens, and are usually located within 1 mm of the pancreas proper.
• This finding is most commonly seen in patients with endocrine neoplasms (43%).
• It may be quite extensive in some individuals.