A 62-year-old woman presented with abdominal discomfort. Imaging studies showed a tumor in the pancreatic tail. At contrast-enhanced CT and macroscopy, the tumor showed cystic, solid and hemorrhagic areas. Histologically, the tumor was well-circumscribed and entirely encapsulated. Some of the tumor cells in the cystic areas were reminiscent of acinar cells, and the majority was arranged in a solid growth pattern. Immunohistochemistry revealed >30% positivity for chymotrypsin, chromogranin A, synaptophysin, and CD56. The diagnosis of a mixed acinar-neuroendocrine carcinoma (MAEC) was made. Review of the English-language literature revealed 44 previously published cases of resected MAECs. We found that, compared to pure acinar cell carcinoma, patients with MAEC have a slightly higher age and are less frequently males, as the male / female ratio was almost equal. The histogenesis of MAEC is still controversial. Due to the small number of cases it is at present not possible to define an evidence-based optimal treatment strategy for these patients.