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Cystic Neuroendocrine Tumors of the Pancreas (c-pNETs): A Single-Center Experience

Vishnu Kommineni1; Maria F. Coakley2; Rahul Pannala3; Longwen Chen3; Jun Zhang4; Nabil Wasif3; Thorvardur R. Halfdanarson4

1University of Alabama Huntsville Regional Campus, Huntsville, AL 35801
2Cork University Hospital, Cork, Ireland
3Mayo Clinic, Scottsdale, AZ 85259
4Mayo Clinic, Rochester, MN 55905

Background: c-pNETs are rare pancreatic neuroendocrine neoplasms which according to some studies are felt to be less aggressive than their solid counterparts.

Methods: We identified all patients diagnosed with a c-pNET at the Mayo Clinic during the period 1990 to 2014. Data on patient characteristics and outcomes of therapy were extracted and analyzed.

Results: 46 patients were identified, 30 males (65%) and 16 females. The median age at diagnosis was 59.5 years (range: 31 to 78 years). 26 (57%) patients had no symptoms at diagnosis. Among symptomatic patients, abdominal pain was the most common symptom. Five patients (12%) had symptoms attributed to hormonal secretion and 4 (9%) had metastases at diagnosis. 10 patients (22%) had a diagnosis of MEN1. The diagnosis was made on a surgical resection specimen in 63%, on a fine-needle aspirate in 26%, a core needle biopsy in 9% and an open biopsy in 2%. Fifteen tumors (33%) were of WHO grade 1, five (11%) of grade 2 and 26 (56%) were ungraded. The majority of patients (66%) had unifocal tumors. Most patients (93%) underwent resection, most often distal pancreatectomy (81%). Most tumors were
located in the tail (52%) followed by the body and head (15% each). The ENETS T-stage was: T1: 30%; T2: 39%; T3: 28%; T4: 2%. Only 4 patients (9%) had nodal involvement. Among resected patients, recurrences occurred in 9 with a median time to recurrence being 68.9 months (range: 6 to 220 months). T-stage did not predict recurrences (p=0.31).

**Conclusion:** c-pNETs are a rare subset of pNETs. They are frequently discovered incidentally, and are usually unifocal, nonfunctional and located in the tail of the pancreas. The prognosis after resection is good with the majority of patients achieving a cure but late recurrences may occur.