

Primary Carcinoid Tumors of the Pancreas: Report of Eight Cases

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Context Primary pancreatic carcinoid tumors (foregut) are very rare. A typical carcinoid syndrome with diarrhea and flushing may be present. The diagnosis is based on the high urinary 5-HIAA (5-hydroxyindole acetic acid) levels (or high serum serotonin levels) or the immunostaining of serotonin (5-HT) in the tumor cells. **Objective** We evaluated clinical presentation, endocrine tumor markers, histology, therapeutic approach and follow up. **Methods** From 1986 to 2011 in our department we observed 211 neuroendocrine (NE) pancreatic tumors and 8 of them (3.8%) were primary carcinoid tumors (5 males and 3 females; averaging 55.8 years, range: 38-69 years). Follow up was updated until December 2012. **Results** Among the eight patients enrolled, 3 were symptomatic. Seven had high serum 5-HT or high urinary 5-HIAA, and one was asymptomatic with immunostaining of 5-HT in tumor cells. Location: 6 body-tail. All were malignant tumors: 7 liver and 1 single nodal

metastases. Markers: 4 high serum 5-HT (up to 176 µmol/L), 7 high urinary 5-HIAA (up to 522 µmol/L). Surgery: 1 left pancreatectomy, 7 biopsy. Histology: 7 NE tumor, 1 negative pancreatic biopsy (liver metastases). Other therapy: 3 treated with somatostatin analogues (SST-A) and chemotherapy (CT), 1 CT and radiometabolic therapy after hepatic artery embolization (HAE), 1 HAE and SST-A, 1 CT. Follow up: 6 dead for disease progression (mean survival 52 months), 2 alive (1 without disease 78 months after surgery; 1 asymptomatic with high 5-HIAA 33 months after SST-A and CT). **Conclusion** Most of primary pancreatic carcinoids are locally advanced tumors or have liver metastases at time of diagnosis, then patients are not amenable to surgery. Although most patients had high 5-HIAA urinary excretion, few patients had carcinoid syndrome. A long term survival may be achieved with multimodal approach, including chemotherapy, in foregut carcinoid tumors.