Primary Carcinoid Tumors of the Pancreas: Report of Eight Cases

Anna C Milanetto¹, Valbona Lico¹, Loredana Iaria², Rita Alaggio², Cosimo Sperti¹, Sergio Pedrazzoli¹, Claudio Pasquali¹

¹Pancreatic and Digestive Endocrine Surgical Unit, Third Surgical Clinic, and ²Pathology; University of Padua. Padua, Italy

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.