We report the case of a 35-year-old female patient with a metastasized carcinoid of the papilla of Vater which is a rare lesion. 96 cases have been published in world literature previously. The carcinoid of the papilla of Vater appears typically as a hormone inactive tumor. It becomes symptomatic by cholestasis and jaundice in most cases and not by carcinoid-syndrome. An association with von Recklinghausen’s disease as described in 25% of cases was not given in our patient. In contrast to the duodenal carcinoid there is no linear relationship between primary tumor size and incidence of metastases. The correct diagnosis was proven by histologic and immunohistochemical methods on specimen taken after endoscopic papillotomy. In spite of sensitive diagnostic methods like endosonography and somatostatin-receptor-scintigraphy exact staging was made intraoperatively in this case. Three months after pylorus pancreatoduodenal resection with lymphadenectomy the patient remained well with no evidence of tumor recurrence.