With an incidence of 0.29/100,000 the malignant forms of neuroendocrine tumors (NET) of the small intestine represent a very rare disease. Recurrence rate of completely resected NET of the small intestine has not clearly been described yet, but depends on the proliferation rate (Ki-67). We report a recurrence in a 64-year-old female with NET of the proximal ileum after 4 years.

A 64-year-old female with family history for colorectal and gastric carcinoma underwent regular screening by her general practitioner. Because of asymptomatic elevation of CA 19-9 a CT-scan was carried out and revealed a tumor of the small intestine of 14 mm in diameter. The tumor was resected and histological workup classified it as a neuroendocrine tumor (pT4, N1, G1, Ki-67 < 1%). After 4 years of follow-up, CT-scan showed two suspicious lesions of the small intestine. Gallium DOTATOC-PET-CT verified tumor recurrence in both sites and segmental resection of the small intestine was performed again. Histological exam of the tumor showed the same histology of NET as before (pT2, N1, G1, Ki-67 < 1%).

Bilocal recurrence of NET of the small intestine with high differentiation and low proliferation rate is rather unlikely with no exact data for recurrence rate in current literature. Following the latest ENETS guidelines of 2009 follow-up should be carried out with regular CT-scan and measurement of chromogranin A according to TNM stage and Ki67 rate at diagnosis. For exact diagnosis and localization somatostatin receptor imaging, either as octreotidescintigraphy or Gallium DOTATOC-PET-CT should be used. As DOTATOC-PET may have higher sensitivity than octreotidescintigraphy, according to latest results it should nowadays be favored. Therefore the two new detected tumors in 2013 could just have been missed by octreotidescintigraphy in 2009.

Even in case of low proliferation index, follow-up for NET should be performed according to current ENET guidelines for at least seven years in order not to miss tumor recurrence. In case of changing diagnostic matters reduced comparability has to be taken in concern.