

Background

Apart from occurring sporadically, insulinoma within the framework of multiple endocrine neoplasia 1 (MEN-1) is well known. The rare presence of multifocal insulinomas has recently been assigned a separate entity (insulinomatosis, see reference). The difficulty of localising insulinomas may be improved by GLP1-receptor imaging.

Case Report

A 48 year old woman had been treated for suspected epileptic seizures for two years (lamotrigine). During another such episode low blood glucose (BG) was detected. During fasting she was unaware of hypoglycaemia (BG 2.3 mmol/L), endogenous hyperinsulinism was established. With a history of treated prolactinoma (operation, quinagolid) and slightly elevated calcium levels MEN-1 was considered.

Operation

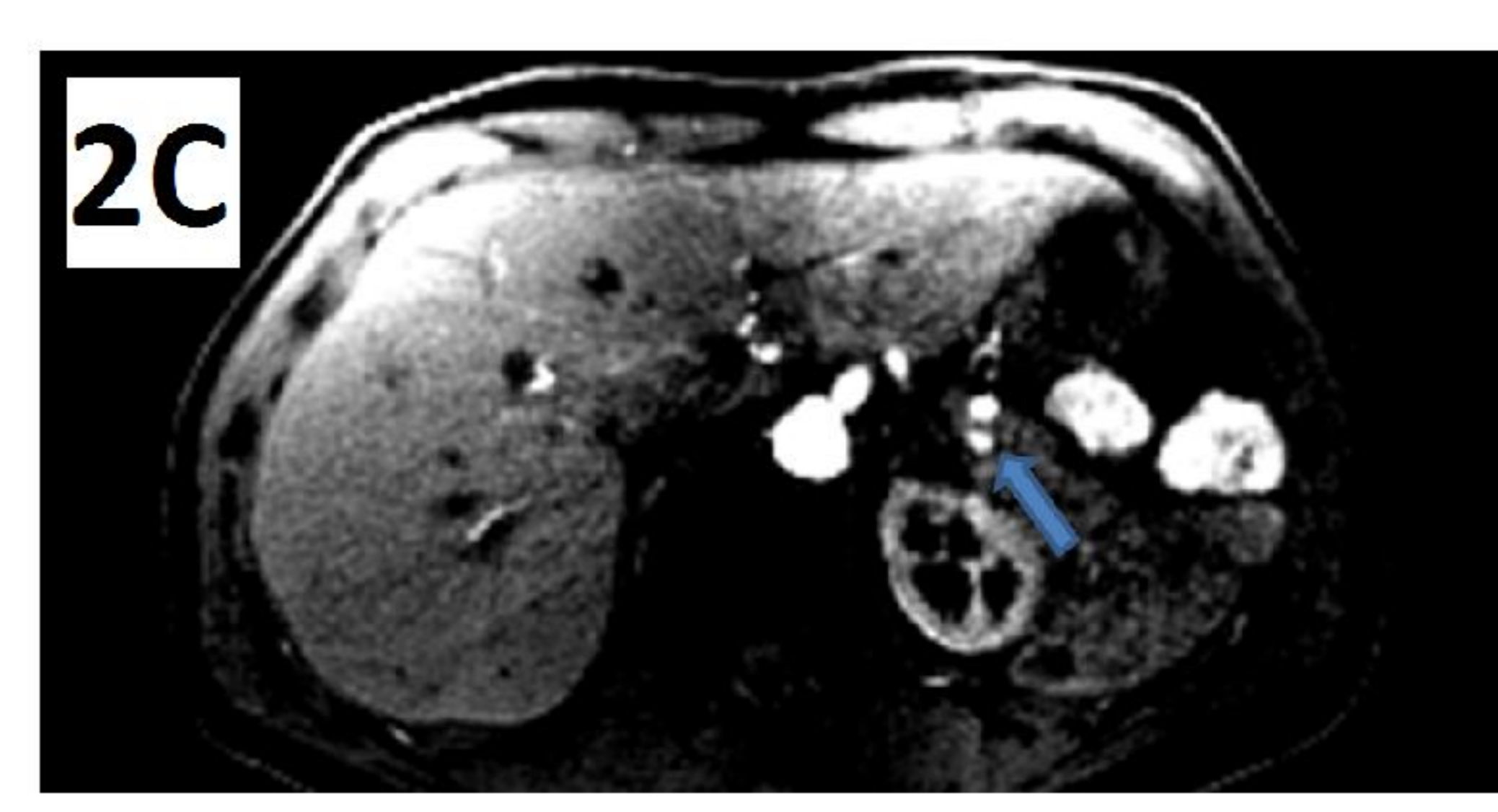
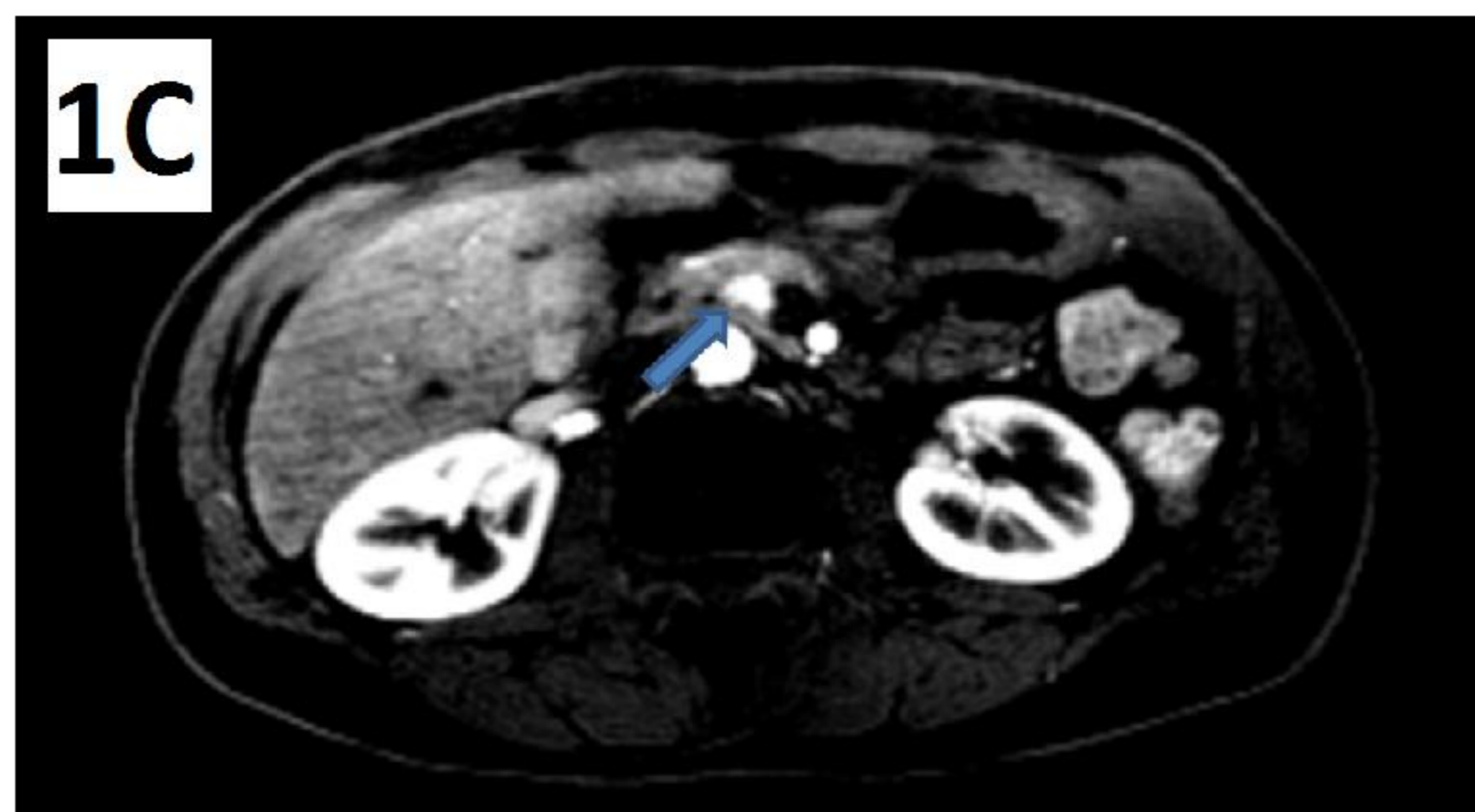
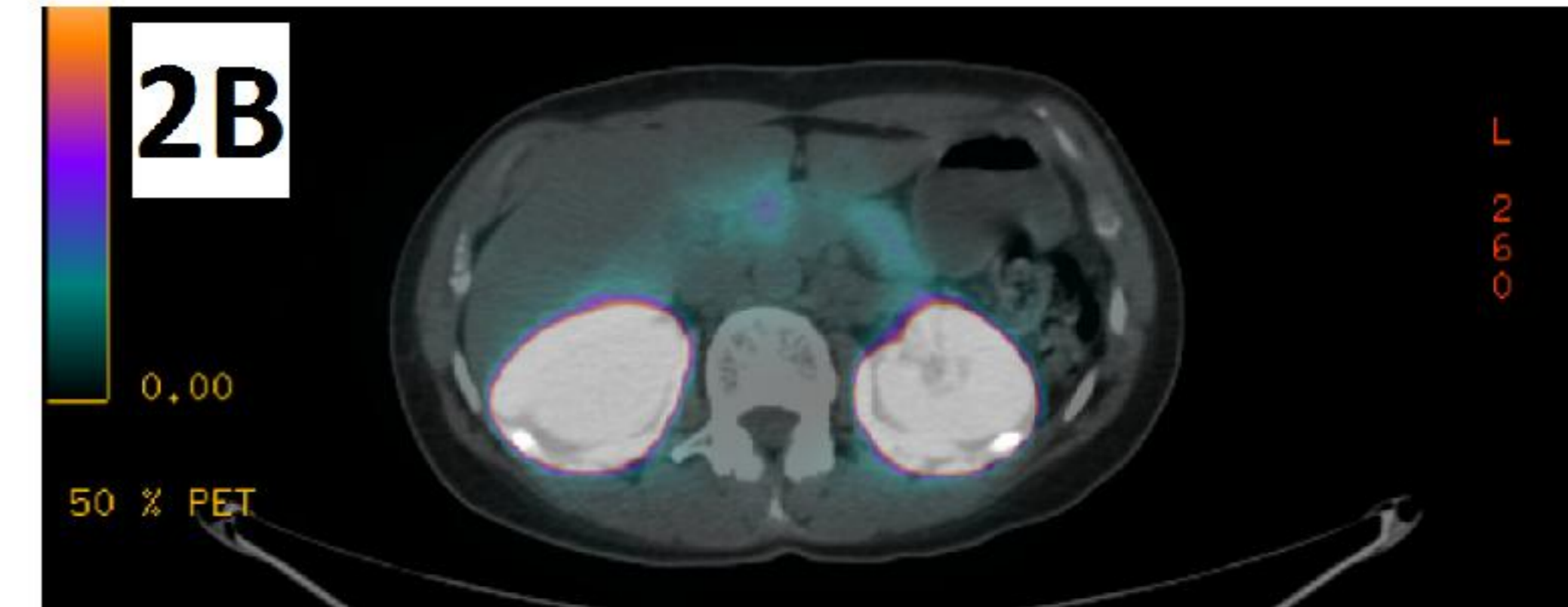
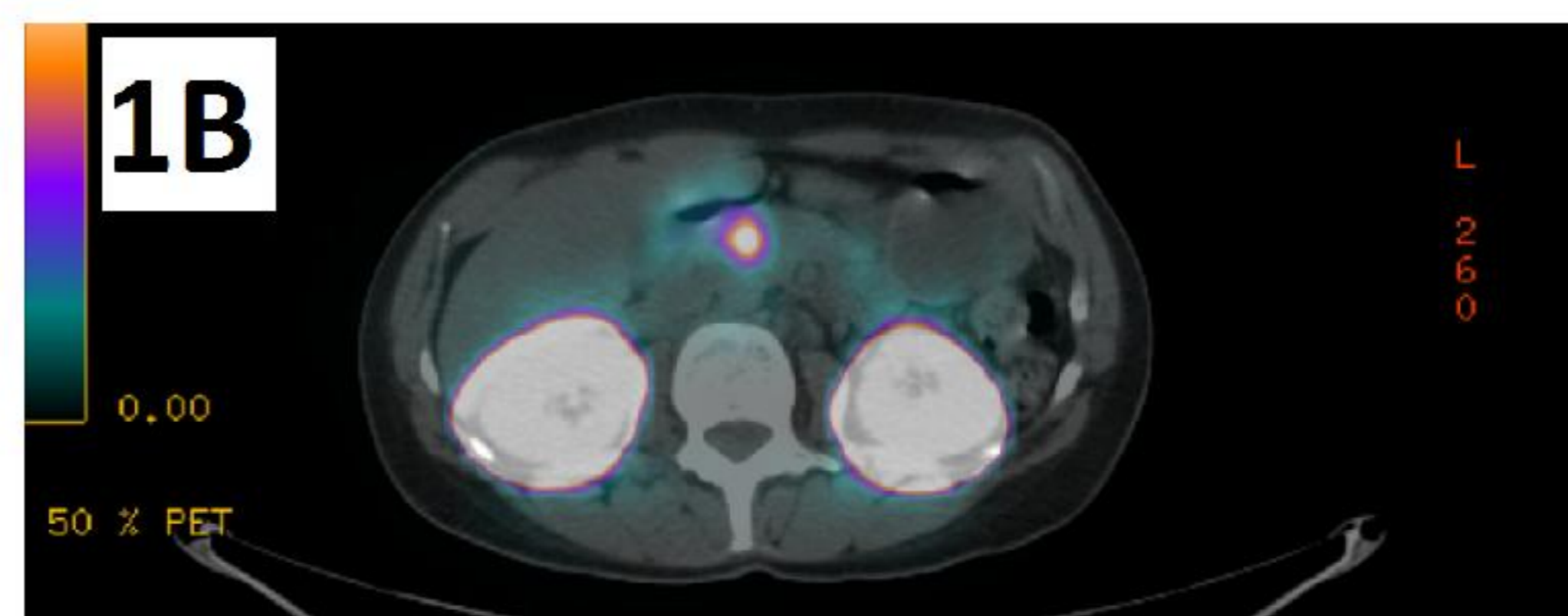
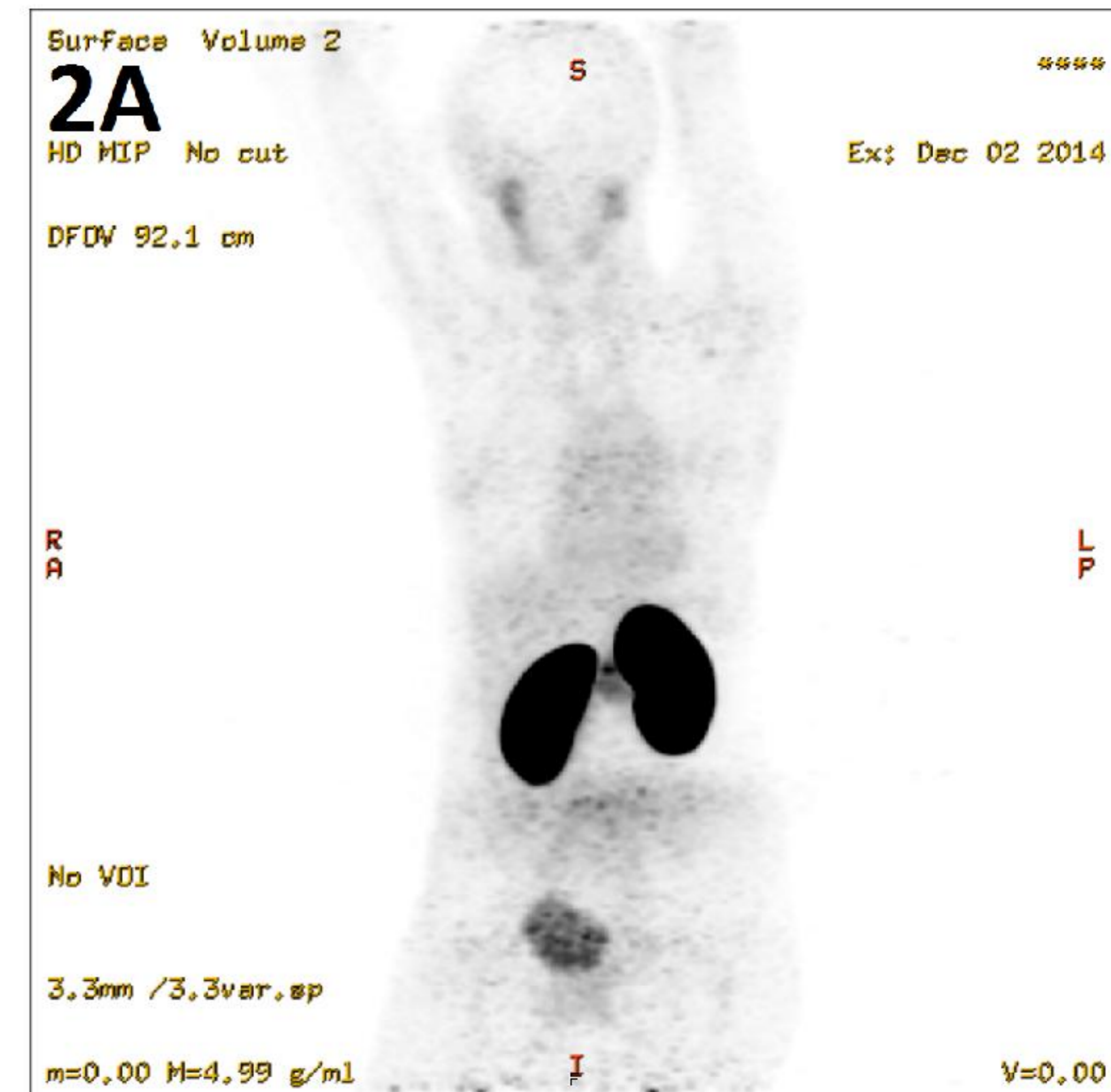
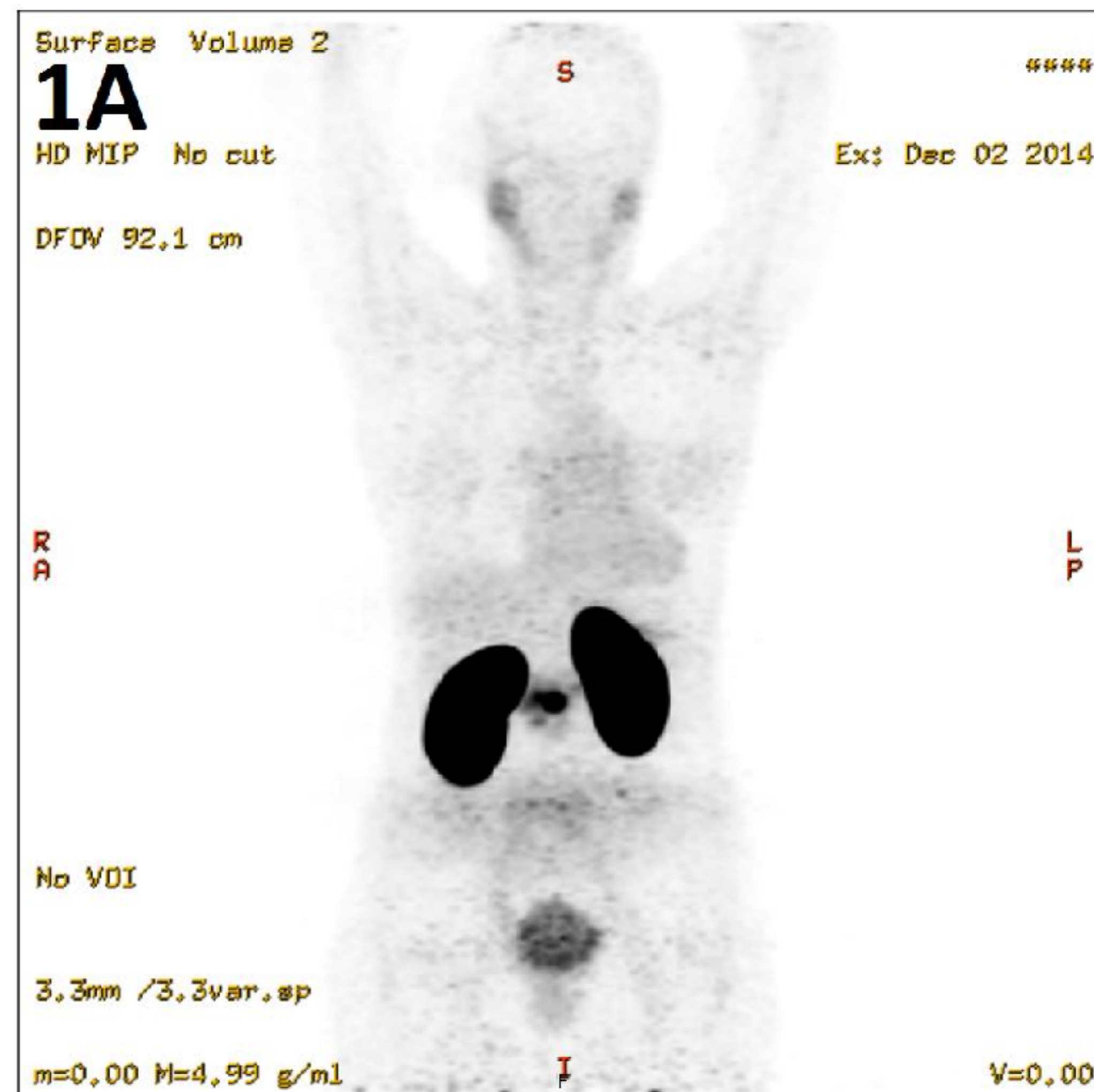
After pretreatment with diazoxide and prednisolone surgery was proposed. Intraoperatively, granular pancreatic tissue was palpated mainly located in the left side up to the head of the pancreas matching with the increased uptake in GLP-1R imaging.

⁶⁸Ga-DOTA-Exendin-4 PET/CT and MRI revealed a major lesion located directly left to the pancreatic head (8 x 13 mm) and smaller lesions in the tail (max. 5 mm); see the figures to the right.

As intraoperative ultrasound did not confirm a focal lesion, left-sided pancreatectomy was performed.

The postoperative course was complicated by a peripancreatic abscess and recurrence of low BG levels requiring diazoxide.

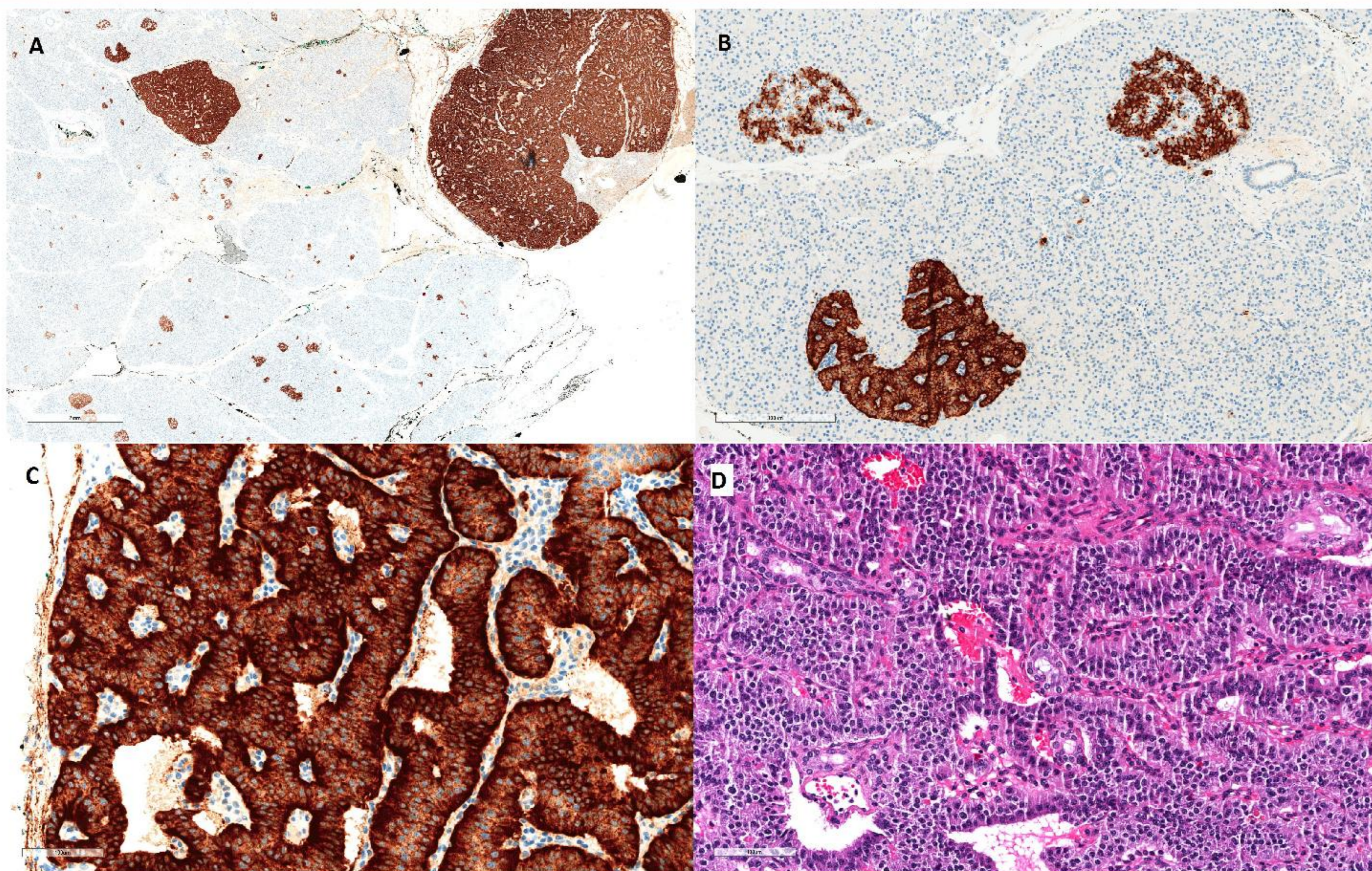
Imaging using ⁶⁸Ga-DOTA-Exendin-4 PET/CT and MR



Left column (1): largest lesion located directly left to the pancreatic head (8 x 13 mm) Right column (2): smaller lesions in the

tail (max. 5 mm); A: ⁶⁸Ga-DOTA-Exendin-4 PET/CT overview; B: cross-section; C MRI (arrows indicating at lesions)

Histopathological details



A: (IHC Insulin) Overview showing 4 microadenomas positive for insulin. B: (IHC Insulin) Microadenoma with trabecular growth compared to two

non-neoplastic islets. C: (IHC Insulin) Detail of biggest microadenoma with trabecular growth. D: (HE) Detail of biggest microadenoma.

Histology / Genetics

Histologically the major lesion proved to be insulin-positive (details to the left), however 37 more small adenoma, mainly with insulin staining were detected establishing the diagnosis of insulinomatosis. Genetic testing for MEN-1 was negative.

Conclusions

1. GLP-R imaging is useful in benign insulinomas and might be useful in the context of MEN-1 in order to separate insulin secreting neuroendocrine tumors (NET) from other secreting and non-secreting NETs,
2. GLP-1R imaging showed positive lesions in this case of insulinomatosis. However, most of the lesions were too small to be detected by ⁶⁸Ga-DOTA-Exendin-4 PET/CT.

Reference

Anlauf, Bauersfeld, Raffel et al. Insulinomatosis. Am J Surg Pathol 2009;33:339-346.

