**Abstract**:

We present a rare case of a 42-year-old man with a long history of von Hippel-Lindau (VHL) disease that was scanned with 68Ga-DOTA-TOC PET/CT for suspicion of disease relapse. 68Ga-DOTA-TOC imaging demonstrated increased DOTA-TOC uptake in pancreas tail and intramedullary and extramedullary spinal hemangioblastomas, only some of which have already been highlighted at MRI examination. This case illustrates the significant role the 68Ga labeled somatostatin receptor analogs PET/CT in the management of the VHL disease.

**References:**

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**Legends:**

**Figure 1**: 68Ga-DOTA-TOC PET/CT. This is a case of a 42-year-old man with von Hippel-Lindau (VHL) disease presenting, at diagnosis in 1984, with retinal haemangiomas (laser-treated), cerebellar and intramedullary spinal hemangioblastomas and bilateral phaeochromocytomas treated with bilateral adrenalectomy. In 1996 a phaeochromocytoma recurrence occurred in the left adrenal region and was surgically resected. In 2004 the patient underwent a pancreaticoduodenectomy for diagnosis of multiple pancreatic NET. In 2016 he was scanned with 68Ga-DOTA-TOC PET/CT for disease follow up. 68Ga-DOTA-TOC PET/CT confirmed recurrence in pancreas tail.

**Figure 2**: 68Ga-DOTA-TOC PET/CT showed also strong uptake in intramedullary (**A**, **B** and **C**) and extramedullary (**D**) spinal hemangioblastomas, only some of which have already been highlighted at previous MRI examination [1, 2]. Two years later, in 2018, a subsequent MRI confirmed the hemangioblastomas already showed up at 68Ga-DOTA-TOC PET/CT in 2016 (**E**).

VHL disease is a dominantly inherited familial cancer syndrome that is associated with a germline mutation of the VHL tumor suppressor gene on the short arm of chromosome 3 (highly penetrant autosomal dominant trait). This disorder is rare, with a prevalence of 1 in 39000–53000. Patients with VHL disease are at risk of developing various benign and malignant tumors, including pancreatic cysts and neuroendocrine tumors, phaeochromocytomas, retinal and central nervous system hemangioblastomas, renal cysts and tumors, endolymphatic sac tumors and reproductive adnexal organs tumors. Treatment of VHL disease is multidisciplinary because of the complexities associated with management of the various types of tumors [3, 4]. Since the prognosis may be improved by an early diagnosis, affected individuals with VHL disease should undergo multimodality imaging, among which seems to play a significant role the 68Ga labeled somatostatin receptor (SSTR) analogs PET/CT [5, 6]. 68Ga-DOTA-TOC PET/CT provide valuable information on the tumor differentiation and expression of somatostatin receptors, and allows for a whole-body evaluation of the disease and essential information for peptide receptor radionuclide therapy planning [7, 8]. 68Ga labeled somatostatin receptor (SSTR) analogs PET/CT could potentially become an integral part in the management of VHL disease.