Complementary role of ¹⁸F-FDG and ⁶⁸Ga-DOTATATE PET/CT in the surveillance of patients with von Hippel-Lindau syndrome

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A 45-year-old woman with a history of von Hippel-Lindau (VHL) syndrome was evaluated during follow up with wholebody positron-emission tomography/computed tomography (PET/CT) scans post administration of ⁶⁸Ga-DOTATATE (targets cell surface overexpression of somatostatin receptors) (Fig. 1A,B) and ¹⁸F-FDG (targets metabolic activity) (Fig. 1C,D) within two consecutive days. 68Ga-DOTATATE was taken up by a cerebral hemangioblastoma (Fig. 1A) and revealed (Fig. 1A,B) 2 lesions in the head and the uncinate process of the pancreas; both were ¹⁸F-FDG negative, indicative of well-differentiated neuroendocrine tumors (NETs). Additionally, ¹⁸F-FDG revealed (Fig. 1C,D) a hypermetabolic tumor adjacent to the left adrenal gland, which was 68Ga-DOTATATE-negative. This tumor was surgically resected and was proven to be an extraadrenal paraganglioma. VHL is a highly penetrant, autosomal, dominantly inherited disorder associated with a germline mutation of the VHL tumor suppressor gene on the short arm of chromosome 3. It is characterized by the development of several benign and malignant tumors, including hemangioblastomas, pancreatic NETs, and paragangliomas [1]. 68Ga-DOTATATE is a radiolabeled somatostatin analog suitable for PET imaging, that has emerged as the imaging standard of reference for the diagnosis of well-differentiated NETs [2,3]. Furthermore, hemangioblastomas, which are highly vascular benign tumors, are known to be 68Ga-DOATATE-positive, as in the present

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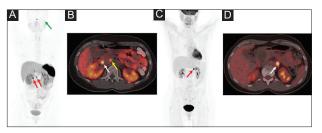


Figure 1 (A) Maximum intensity projection (MIP) ⁶⁸Ga-DOTATATE positron-emission tomography (PET) image of the head and torso, showing a focal lesion in the left cerebellum (green arrow), corresponding to a known hemangioblastoma, and 2 foci of abnormally increased radiotracer uptake in the abdomen (red arrows). (B) Axial fused ⁶⁸Ga-DOTATATE PET/CT image of the abdomen, showing (white arrow) a focal lesion in the pancreatic head (SUVmax: 41.7) and a second lesion (yellow arrow) in the uncinate process of the pancreas (SUVmax: 32.7), indicative of neuroendocrine tumors. (C) MIP ¹⁸F-FDG PET image of the head and torso, showing a focal, intensely hypermetabolic lesion in the left upper abdomen (red arrow). (D) Axial fused ¹⁸F-FDG PET/CT image of the abdomen, showing (white arrow) ¹⁸F-FDG avid lesion (SUVmax: 20.3) located below the pancreatic tail, adjacent to the left adrenal gland, and histology revealed extra-adrenal paraganglioma

case [2,4]. In addition, ¹⁸F-FDG is the most widely used PETtracer for oncologic purposes, as it can contribute significantly to the detection and follow up of various VHL-related tumors. The presented data highlights the complementary role of ⁶⁸Ga-DOTATATE and ¹⁸F-FDG for the accurate diagnosis of uncertain lesions in the context of the surveillance for patients with VHL.

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