A rare adult renal neuroblastoma better imaged by ^{18}F-FDG than by ^{68}Ga-dotanoc in the PET/CT scan

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Abstract
Primary renal neuroblastoma is an uncommon tumor in children and extremely rare in adults. We present a case of a middle aged female having a large retroperitoneal mass involving the right kidney with features of neuroblastoma on pre-operative histopathology. Whole-body fluorine-18-fluoro-deoxyglucose positron emission tomography (^{18}F-FDG PET/CT) and ^{68}Ga-dotanoc PET/CT scans performed for staging and therapeutic potential revealed a tracer avid mass replacing the right kidney and also pelvic lymph nodes. The ^{18}F-FDG PET/CT scan showed better both the primary lesion and the metastases in the pelvic lymph nodes than the ^{68}Ga-dotanoc scan supporting diagnosis and treatment planning.

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Introduction

In this case we discuss the features of metabolic imaging in an adult with a very rare renal neuroblastoma. Neuroblastoma is the most common extracranial solid malignancy in paediatric age accounting to: 8% of all childhood cancers with an annual incidence of 10.5 cases per million children. Its annual incidence in adulthood is only 0.12 to 0.2 per million population. The adult renal neuroblastoma is extremely rare and only six such cases have been published so far to the best of our knowledge [1-6]. The metastatic potential of neuroblastoma at diagnosis is quite high involving mainly bone and bone marrow (55% and 70% respectively) [7]. Imaging studies support diagnosis and staging of the disease. We report here a case of a 53 years old female examined by hybrid ^{18}F-FDG PET/CT (GE Discovery 710, GE Healthcare, Milwaukee, WI, USA) and by ^{68}Ga-dotanoc PET/CT. ^{68}Ga-dotanoc is a short peptide analogue to somatostatin (Nal3-octreotide-NOC) linked to the positron emitter ^{68}Ga by a bifunctional chelate-1,4,7,10-tetraazacyclododecane-1,4,7,10-tetraacetic acid (DOTA).

Case Report

A 53 years old female farmer post-treated for left breast carcinoma presented with a slow growing, painless tumour on the right renal region and hypertension of 144/92 mmHg of 2 months duration. The ultrasonography of the abdomen revealed a large retroperitoneal mass involving the right kidney. The histopathology and immunohistochemistry of the biopsied mass lesion showed tumor cells arranged in diffuse sheets with occasional pseudorosetting. Individual tumor cells were relatively monomorphic with higher N:C ratio (N:C is nuclear-cytoplasmic ratio and its normal value for the kidney is ~2:1), hyperchromatic nucleus and scant amount cytoplasm. The tumor cells showed strong membranous positivity for CD99, CD 56 and cytoplasmic positivity for NSE and synaptophysin, diagnostic of neuroblastoma. Urinary catecholamine levels (vanillylmandelic acid and homovanillic acid) were within normal limits. The whole-body ^{68}Ga-dotanoc PET/CT followed by ^{18}F-FDG PET/CT scan were done for staging, prognostication and to find out whether radionuclide therapy was feasible. The maximum intensity projection (MIP) image of ^{68}Ga-dotanoc PET/CT (Figure 1a) revealed in the right renal region moderate inhomogenous tracer uptake (arrow) with tracer avid pelvic lymph

nodes (broken-arrow), while MIP image of ^{18}F-FDG PET/CT (Figure 1b) showed in the right renal region intense tracer uptake (arrow) and multiple pelvic lymph nodes (broken-arrow). The coronal and transaxial CT, fused ^{68}Ga-dotanoc PET/CT and ^{18}F-FDG PET/CT images showed a non-enhancing lobulated soft tissue mass (~12.6x18.6x11.5cm) in the right renal fossa almost replacing the right kidney with tracer uptake, with standardised maximum uptake value (SUV max~5.9) for dotanoc and with SUVmax~18.2 for ^{18}F-FDG. The ^{18} F-FDG PET/CT scan showed tracer avid pelvic lymph-nodes (Figures 1c-k). The right adrenal gland was normal and visualized separately from the kidney. The patient underwent right nephrectomy and lymph nodes dissection. The adrenal gland appeared to be uninvolved and was not dissected. We also evaluated the screening score for this patient. The SUVmax calculated from the ^{68}Ga-dotanoc scan of the primary disease was low not favouring the peptide receptor radionuclide therapy, and so surgery was the best available option.

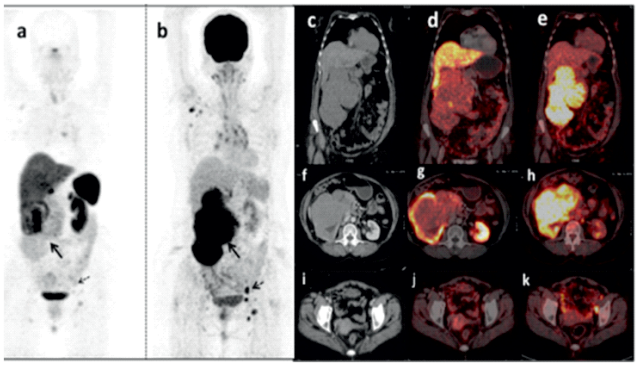

Figure 1. The maximum intensity projection (MIP) image of ^{68}Ga-dotanoc (Figure 1a) revealed moderate inhomogenous tracer uptake in the right renal region (arrow) with mild tracer avid pelvic lymph nodes (broken-arrow), while MIP image of ^{18}F-FDG (Figure 1b) showed intensed tracer uptake in the right renal region (arrow) and multiple pelvic lymph nodes (broken-arrow). Coronal and transaxial PET/CT and fused images (Figure 1c-k) showed tracer avid (SUVmax~5.9 in dotanoc) and SUVmax~18.2 in ^{18}F-FDG PET images of a non-enhancing lobulated soft tissue mass (~12.6x18.6x11.5cm) in the renal fossa almost replacing the right kidney with tracer avid pelvic lymph nodes.

Discussion

Less than 30 cases of primary renal neuroblastoma in paediatric population have been reported so far, but primary renal neuroblastoma in adults is rare [8, 9]. Fan R. (2012) reviewed 8 cases of primary renal neuroblastoma in paediatric patients collected over 15 years (largest series so far) and found: male predominance, typical presentation of the neuroblastoma as an abdominal mass, with hypertension with or without elevated urinary catecholamines level, and an average age of 17 months (range 3-39 months) [10].

It is considered that renal neuroblastoma originates from the neural crest tissue in the renal hilum/pelvis. The hypothesis for its renal origin is supported by the entrapped adrenal medullary tissue in the kidney as a developmental remnant or from the sympathetic ganglion within the kidneys [11]. The adrenal glands are usually not involved in renal neuroblastoma. Furthermore, the characteristic calcification has been shown to be absent in renal neuroblastoma resembling nephroblastoma which also shows absent calcification. Imaging cannot differentiate between nephroblastoma and neuroblastoma and histopathology is required for diagnosis [10, 12]. In our case the fluorodeoxyglucose and the somatostatin receptors in PET/CT imaging revealed differential avidity but without any calcified focus within the tumor and an uninvolved right adrenal gland.

Renal neuroblastoma commonly manifests at a higher stage at the time of diagnosis and is considered aggressive in nature. Imaging is usually done at first to diagnose the metastatic sites, in bone marrow and bones followed by liver and lymph nodes metastases [7]. Functional metabolic imaging using the glycolytic pathway and somatostatin receptors based imaging have shown in neuroblastoma patients encouraging results in staging, prognostication and the potential for therapeutic strategies. Fluorine-18-FDG imaging has been found to better define the extent of the disease also in post-operative recurrence cases. Though histopathological diagnosis was necessary in this case, functional imaging has been very useful [12-15].

In conclusion: In this rare case of adult renal neuroblastoma the ^{18}F-FDG PET/CT scan showed better both the primary lesion pelvic lymph nodes and metastases than the ^{68}Ga-dotanoc scan supporting diagnosis and treatment planning.

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