Case Report

F-18 fluorodeoxyglucose positron emission tomography "super scan" in a patient of metastatic primitive neuroectodermal tumor of the kidney

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ABSTRACT

We report F-18 fluorodeoxyglucose (FDG) "positron emission tomography (PET) super scan" akin to "super scan" of conventional skeletal scintigraphy, in a rare case of primitive neuroectodermal tumor (PNET) of the kidney. A twelve year old male patient of metastatic PNET of the kidney was subjected to a "true" whole body F-18 FDG PET scan including lower limbs and skull region as per the institution protocol. The images revealed extensive hypermetabolic areas corresponding to the computed tomography described renal, hepatic, and pancreatic lesions along with intense and non-uniform uptake in the marrows of axial and appendicular skeletal system. Interestingly, low background tracer concentration was observed along with very low F-18 FDG uptake in the brain, skeletal muscles of limb, mediastinum, and bowel. In view of these findings, the scan can be interpreted as "PET super scan" due to its resemblance with the super scan of skeletal scintigraphy. A repeat F-18 FDG PET scan after chemotherapy revealed marked treatment response with disappearance of "super scan"-like pattern, reduction in number, size, metabolic activity of the lesions, and stimulated marrow sans the previously diseased portion. Though uncommon, the reporting physician should be aware of "PET super scan" and its implications as described in this case.

Keywords: F-18 fluorodeoxyglucose positron emission tomography, marrow "flip flop", primitive neuroectodermal tumor of kidney, super scan

INTRODUCTION

"Super scan" is a well described phenomenon on skeletal scintigraphy which is characterized by high skeletal to soft tissue ratio, uniform symmetrically increased bone uptake, and absent renal visualization.[1] Sye, *et al.* hypothesized that diseased bone shows increased uptake of radiopharmaceutical leading to reduced phosphate excretion and production of faint renal images on bone scan.[2] We report a case of F-18 fluorodeoxyglucose (FDG) positron emission tomography (PET) super scan which shows features akin to super scan of skeletal scintigraphy and resolution of these features following initial phase of chemotherapy.

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CASE REPORT

A twelve-year old male presented with multiple joint pains, periorbital and sternal swellings of one-month duration. Contrast-enhanced computed tomography (CT) scan of the abdomen revealed poorly enhancing, hypodense bilateral renal masses ranging from 4 cm to 5 cm in size along with multiple hepatic and pancreatic space occupying lesions. A 4.3-cm exophytic mass was noted in the lower pole of right kidney [Figure 1]. CT-guided biopsy from this mass revealed feature of renal primitive neuroectodermal tumor (PNET)/ Ewing's sarcoma [Figure 2].

Thereafter, patient underwent a "true" whole body PET scan including bilateral limbs and skull region as per the standard institution protocol for pre-treatment staging of the disease. The

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images revealed extensive hypermetabolic areas in CT described renal (SUV_{max}: 6.9), hepatic (SUV_{max}: 7.2), and pancreatic lesions (SUV_{max}: 5.4). In addition, intense and non-uniform uptake was seen in marrows of axial and appendicular skeletal system with distinct focus in palpable sternal swelling (SUV_{max}: 4.5). A medium-sized focus was also seen in left posterior chest wall with extension into basal pleura of left lung. Interestingly, low background tracer concentration was observed along with very low F-18 FDG uptake in the brain (SUV_{max} of 3.3 in occipital region and 2.9 in rest of the brain), skeletal muscles of limb, mediastinum, and bowel. Due to its resemblance with super scan of skeletal scintigraphy, these scan findings can be termed to represent "PET super scan" [Figure 3].

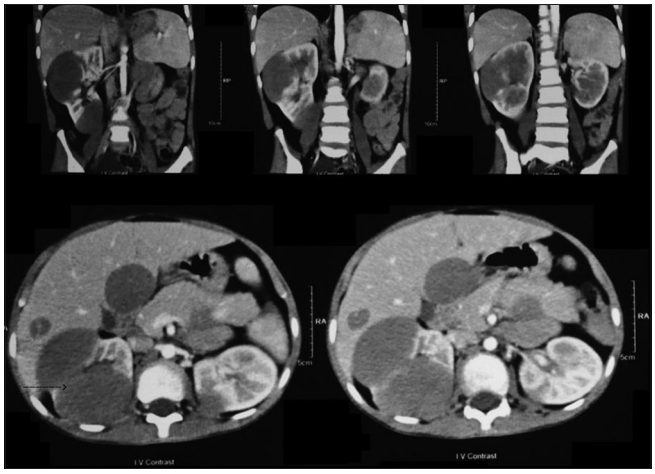

Figure 1: Contrast-enhanced computed tomography scan of the abdomen revealed poorly enhancing, hypodense bilateral renal masses ranging from 4 cm to 5 cm in size along with multiple hepatic and pancreatic space occupying lesions. A 4.3-cm exophytic mass was noted in the lower pole of right kidney

Patient was subjected to six cycles of Euro-Ewing 99 protocol including Vincristine, Ifosfamide, Doxorubicin, and Etoposide chemotherapeutic regimen as per the institution protocol.[3] Repeat PET scans were done after three and six cycles, latter being done after 4 months of the initial scan [Figure 4a]. The images revealed marked treatment response with disappearance of "super scan"-like pattern, reduction in number, size, and metabolic activity of the lesions that were shown in the earlier scan. SUV_{max} of the occipital region of the brain increased from the previous value of 3.3 to 9.9. For rest of the brain, the SUV_{max} increased from 2.9 to 8.6 [Figure 4b]. The comparison of femoral marrow activity in the pre- and post-treatment scans showed hypermetabolic areas of stimulation in post-treatment scan, which is clearly missing from the previously diseased marrow and GCSF-induced stimulated marrow being seen in

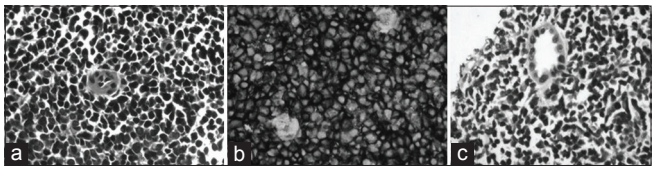

Figure 2: Histology of renal biopsy specimen from the lesion (H and E staining, ×10) showing sheets of malignant small round blue cells. (a) On immunohistochemistry, the tumor cells show strong and complete membranous positivity for Mic-2. (b) They were immuno-negative for WT-1. (c) Tumor cells were also negative for other markers in the round cell panel namely cytokeratin, Leukocyte-common antigen, desmin, and synaptophysin (not shown here). The findings are those of primitive neuroectodermal tumor/Ewing's sarcoma of the kidney

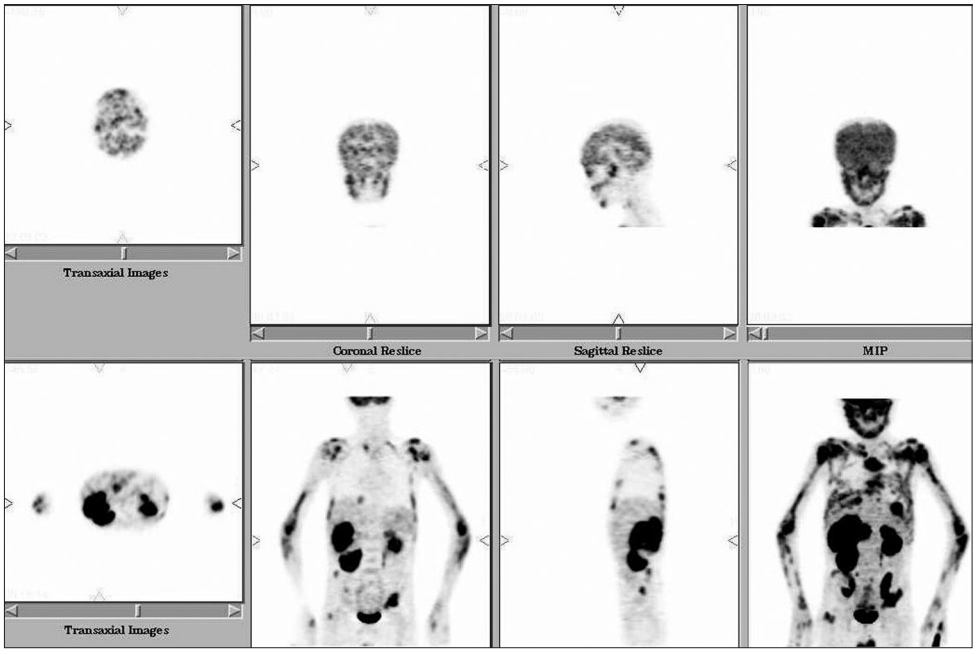

Figure 3: "True" whole body PET scan including bilateral limbs and skull region shows extensive hypermetabolic areas in computed tomography described renal (SUV_{max}: 6.9), hepatic (SUV_{max}: 7.2), and pancreatic lesions (SUV_{max}: 5.4). Intense and non-uniform uptake was seen in marrows of axial and appendicular skeletal system with distinct focus in palpable sternal swelling (SUV_{max}: 4.5) and in left posterior chest wall with extension into basal pleura of left lung. Scan shows low background tracer concentration with very low F-18 FDG uptake in the brain (SUV_{max} of 3.3 in occipital region and 2.9 in rest of the brain), skeletal muscles of limb, mediastinum, and bowel. Due to its resemblance with super scan of skeletal scintigraphy, these scan findings can be termed to represent "PET super scan"

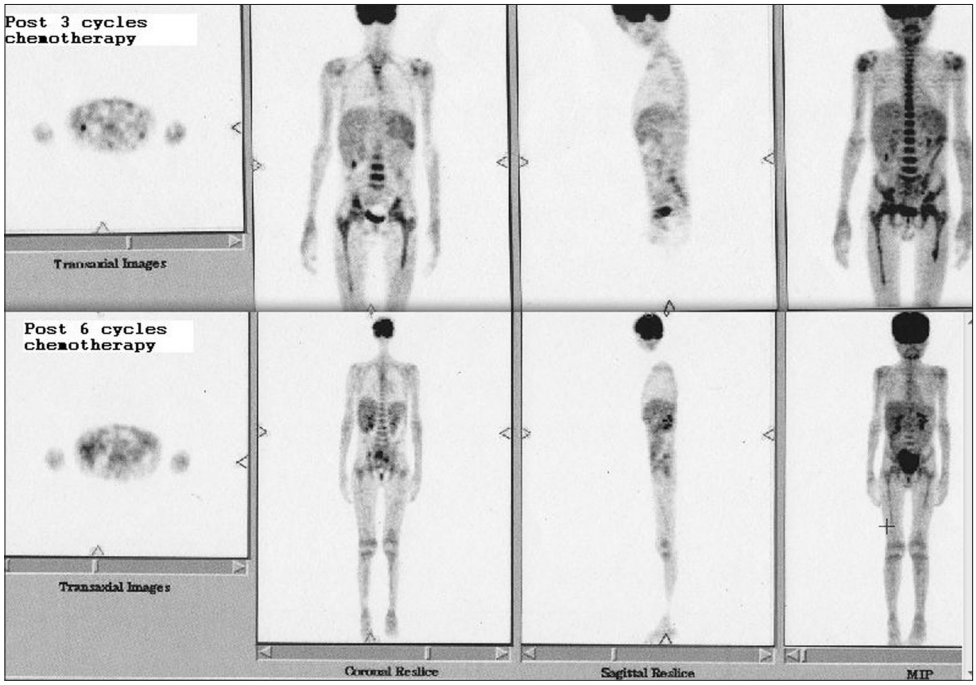

Figure 4a: Repeat PET scans were done after three and six cycles, latter being done after 4 months of the initial scan

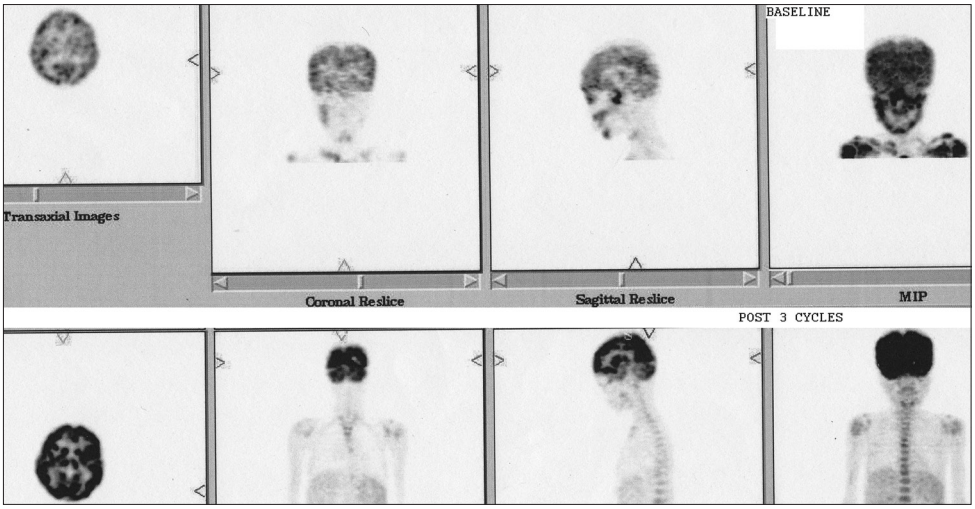

Figure 4b: The images show marked treatment response with disappearance of "super scan"-like pattern, reduction in number, size, and metabolic activity of the lesions that were shown in the earlier scan. SUV_{max} of the occipital region of the brain increased from the previous value of 3.3 to 9.9. For rest of the brain, the SUV_{max} increased from 2.9 to 8.6

previously uninvolved marrow. We have termed this pattern as "marrow flip flop" and it appears that areas showing marrow flip flop are the ones that demonstrate treatment response [Figure 4c].

DISCUSSION

PNET of the kidney was first reported by Mor, *et al.*[4] It is an extremely rare malignant tumor with fewer than 50 cases in English literature. Renal PNET is still rarer in children and adolescents age group with only four cases being reported in the pediatric age group.[5-7] To the best of our knowledge, we are reporting fifth such case. It usually presents in advance stage, behaves more aggressively than PNET at other sites, and usually shows poor response to the chemotherapy.[8]

Similar characteristics of diffuse and intense hypermetabolism throughout the skeleton have been described in the earlier publications.[9-11] However, the scan in this case shows a very high contrast between the metastatic and non-metastatic organs with extremely low F-18 FDG uptake in brain, muscles of limbs, mediastinum, and bowel. Unique feature of this case is demonstration of reversal of aforementioned F-18 FDG avidity after initial chemotherapy in the follow-up scan. Another unusual feature of this case is the excellent chemotherapeutic response shown in this patient; PNET of kidney is known

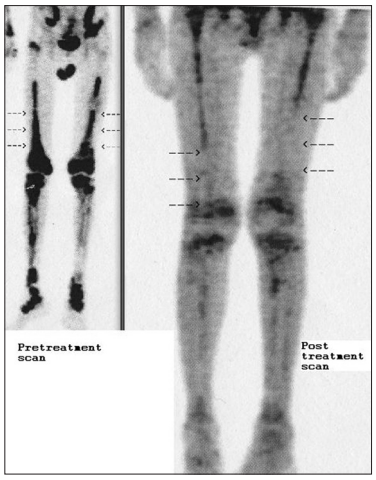

Figure 4c: The comparison of femoral marrow activity in the pre- and post-treatment scans shows hypermetabolic areas of stimulation in post-treatment scan, which is clearly missing from the previously diseased marrow and G-CSF-induced stimulated marrow being seen in previously uninvolved marrow termed as "marrow flip flop"

to respond poorly to chemotherapy and a functional imaging modality may serve as a useful tool to demonstrate early response in such rare cases. "Marrow flip flop" could be due to involved marrow being replaced by fibrosis as a treatment response and hence would not demonstrate the marrow stimulation effect of colony-stimulating factors on a PET scan done to assess therapeutic response.

In conclusion, though uncommon, the reporting physician should be aware of findings of "PET super scan" as demonstrated in this patient. Reversal of these abnormalities on functional imaging following therapy guides the clinician to choose the best available regimen for this otherwise chemotherapy naïve disease.

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