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Case Report

^{18}Fluorine-fluorodeoxyglucose PET-CT findings in a case of rarely seen metastatic adult extrarenal Wilms' tumor of retroperitoneum presenting as lower limb edema

ABSTRACT

Wilms' tumor also called as nephroblastoma is commonly seen extracranial solid tumor involving kidneys in children. Rarely, Wilms' tumor can arise from ectopic nephrogenic remnants located outside the kidneys. Extrarenal Wilms' tumor comprises 3% of total Wilms' tumor with its incidence even less common in adults. We report the staging and restaging fluorodeoxyglucose positron emission tomography-computed tomography findings in a case of locally advanced metastatic extrarenal adult Wilms' tumor involving the retroperitoneum.

Keywords: Adult extrarenal Wilms' tumor, lower limb edema, national Wilms' tumor study, retroperitoneum

INTRODUCTION

Adult Extrarenal Wilm's Tumor (ERWT) refers to the presence of nephroblastic tumor outside the kidneys in a patient with normal bilateral kidneys with age >15 years.[1] Extrarenal nephroblastoma was first described by Moyson *et al*. in 1961 in a patient with mediastinal mass.[2] Thereafter, the reported incidence of ERWT was seen to be approximately 0.2 per million per year with adult ERWT even rarely reported in literature.[3,4]

CASE REPORT

A 36-year-old male patient presented with a chief complaint of lower limb edema associated with vague abdominal pain. Clinical examination of the abdomen was grossly unremarkable. USG abdomen with venous Doppler was suggestive of heteroechoic mass in the retroperitoneum with associated thrombus in the left renal vein. Whole-body ^{18}fluorine-fluorodeoxyglucose (FDG) positron emission tomography-computed tomography (PET-CT) [Figure 1a] done for further characterization shows an area of increased tracer uptake in the left lumbar region (black arrow). Axial PET, CT, and fused PET-CT images localize the increased tracer uptake in the abdomen to metabolically avid heterogeneously enhancing solid-cystic soft-tissue mass [white solid arrow; Figure 1b-d] in left-sided retroperitoneum. The mass is seen to invade the left renal vein superiorly with FDG-avid tumor thrombus [white dashed arrow; Figure 1e-g] extending to infrahepatic inferior vena cava. Non-FDG-avid random lung nodule [white dotted arrow; Figure 1h-j] was also noted. In view of locally aggressive FDG-avid solid cystic retroperitoneal lesion, a provisional diagnosis of extragonadal germ cell tumor was considered. However, workup with serum beta-human

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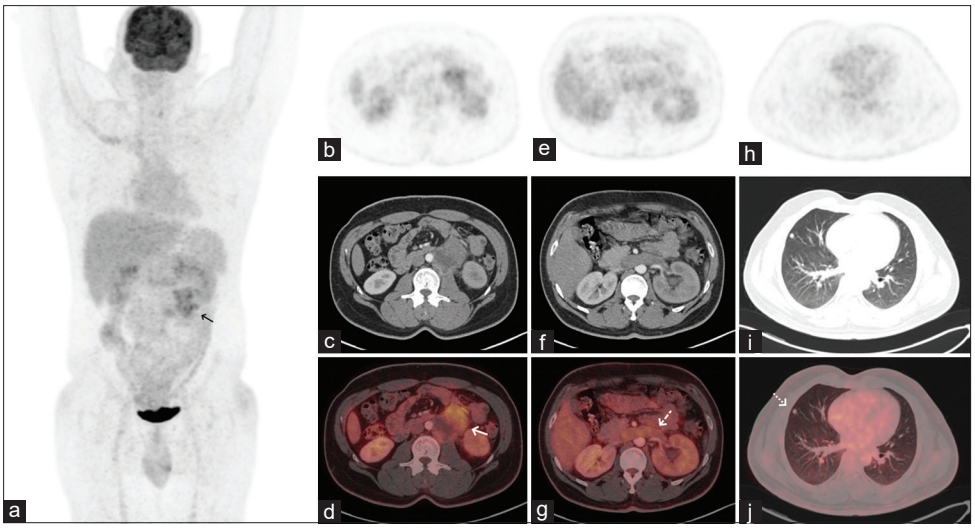

Figure 1: Whole-body fluorodeoxyglucose positron emission tomography MIP image (a) reveal increased tracer uptake in the left lumbar region (black arrow). Transaxial positron emission tomography, computed tomography, and fused positron emission tomography-computed tomography images localize the increased tracer uptake to fluorodeoxyglucose-avid solid-cystic soft-tissue mass (white solid arrow; b-d) in left-sided retroperitoneum invading left renal vein (e-g: white dashed arrow; g) along with random right lung nodule (h-j) :white dotted arrow; j)

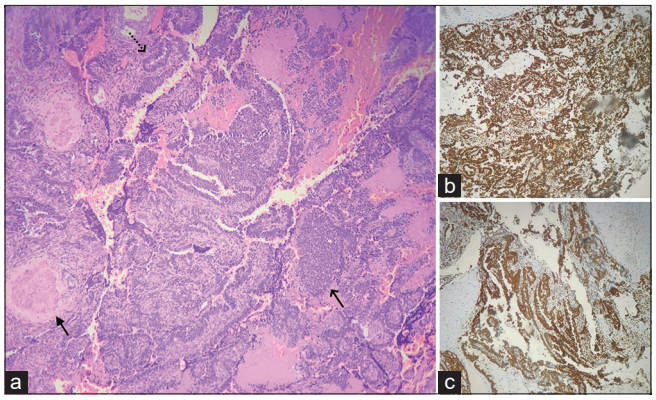

Figure 2: Microphotograph (a; H and E stain, original magnification × 10) of sections from soft-tissue mass shows mitotically active primitive tumor cells (solid arrow) arranged as tubules (dashed arrow) at places amidst a loose spindle cell mesenchymal stroma (solid filled arrow). Immunohistochemistry shows a positive expression of WT1 protein in the primitive cells and tubules (b) along with PAX8 expression in the renal tubules (c)

chorionic gonadotropin and alpha-fetoprotein was normal. Histopathological examination [Figure 2a] from the soft-tissue mass shows primitive mitotically active tumor cells (solid arrow) arranged as tubules (dashed arrow) at places amidst a loose spindle cell mesenchymal stroma (solid-filled arrow). Furthermore, immunohistochemistry shows a positive expression of WT1 protein in the primitive cells and tubules [Figure 2b] along with PAX8 expression in the renal tubules [Figure 2c]. The presence of histological features of blastemal, epithelial, and stromal components associated with the phenotypic expression of WT1 protein and PAX8 in the tubules in a patient aged >15 years excludes teratoma and confirms a diagnosis of adult triphasic Wilms' tumor. In view of locally advanced inoperable adult extrarenal Wilms' tumor (ERWT) with metastatic lung nodule, Stage IV adult ERWT was considered according to national Wilms' tumor study (NWTS) staging, and NWTS-5 chemotherapy drug regimen (Doxorubicin, vincristine, cyclophosphamide, and etoposide) was administered. Follow-up PET-CT [Figure 3a and b] done for response assessment shows near-total resolution of FDG-avid focus in the left-sided abdomen. Staging and restaging transaxial fused FDG PET-CT images show significant interval reduction in the metabolic activity of left-sided retroperitoneal lesion associated with increase in the necrotic component [Figure 3c and d SUVmaxbw - 6.8 vs. 2.4; white solid arrow). Transaxial staging and restaging fused FDG PET-CT images also show interval reduction in the metabolic activity of thrombus [Figure 3e and f SUVmaxbw - 3.8 vs. 2.2; white dashed arrow] along with total resolution of lung nodule [Figure 3g and h].

DISCUSSION

ERWT most likely develops from oncogenic mutation of the ectopic nephrogenic cell rests along the craniocaudal migratory pathway of primitive metanephros and mesonephros. Association of ERWT with horseshoe-shaped kidney, spinal dysraphism, and presence of WT1 gene mutations in 25% of these tumors supports the abovementioned hypothesis.[5,6] The various reported sites of EWRT include mediastinum, retroperitoneum, inguinal canal, lumbosacral region, and pelvis with presenting complaints mainly due to the local pressure effects of the tumor in the involved region.[7,8] Even though extragonadal germ cell tumor can present as isolated solid cystic mass in the retroperitoneum, the presence of renal vein invasion

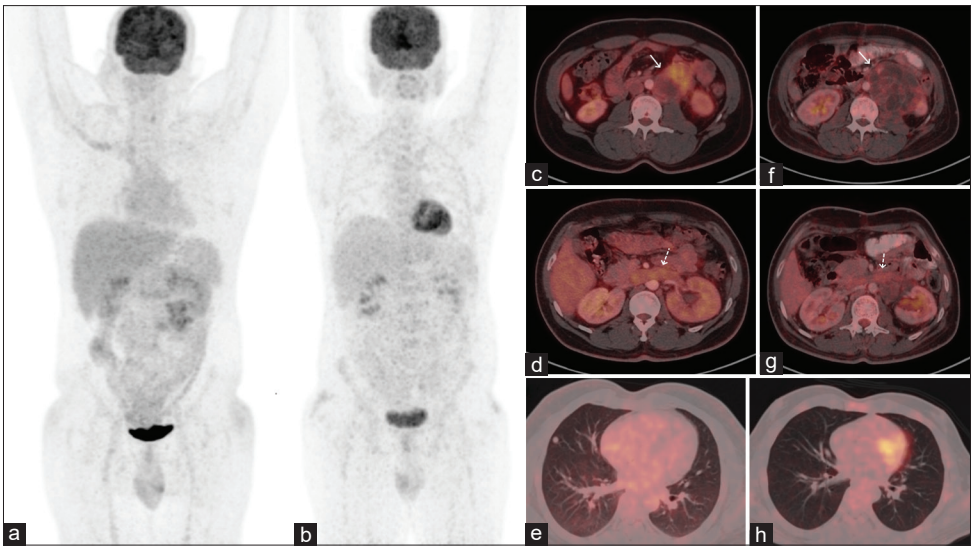

Figure 3: Restaging fluorodeoxyglucose positron emission tomography-computed tomography (a and b MIP) done for response assessment shows near-total resolution of fluorodeoxyglucose-avid focus in the left-sided abdomen. Transaxial images show significant interval reduction in the metabolic activity of left-sided retroperitoneal lesion (c and d; white solid arrows; SUVmax - 6.8 vs. 2.4), and tumor thrombus (e and f; white dashed arrows; SUVmax - 3.8 vs. 2.2) along with the total resolution of lung nodule (g and h)

and thrombus without elevation of tumor markers should prompt the possibility of this rare entity as seen in this case. As seen with childhood intrarenal Wilms' tumor, adult ERWT also invades renal vein with similar morphological, behavioral, and histopathological features.[9] In view of the paucity of literature, staging and management protocol guidelines given by NWTS for childhood Wilms' tumor are even followed for adult Wilms' tumor in most of the centers, leading to variable treatment response.[9] Wilms' tumor of kidney and childhood ERWT are usually FDG avid and PET plays an important role in tumor staging, excluding histologically active residual disease following chemotherapy and diagnosing early recurrence of tumor.[10,11] Utility of FDG PET in the management of adult ERWT is less commonly reported in literature.

CONCLUSION

Based on the image findings in this index patient, FDG PET-CT can be used for staging and response assessment in these rarely seen tumors.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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