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Case Report

An Unusual Case of Primary Renal Primitive Neuroectodermal Tumor in a Middle-Aged Man

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ABSTRACT

PNET of the kidney is a very rare tumor and extension to the IVC is rarer. We report a case of primitive neuroectodermal tumor(PNET) of the kidney with IVC thrombus in a 38 year old man. The patient was managed with radical nephrectomy with IVC thrombectomy and post operative chemotherapy. The patient is recurrence free at one year of follow up.

Key-words: Kidney, IVC Thrombus, Primitive Neuroectodermal tumour

INTRODUCTION

Primitive neuroectodermal tumors (PNET) are a group of small round cell malignancies with neural crest origin. They can be central or peripheral in origin, peripheral origin is extremely rare out of which genitourinary PNET are the rarest. Renal PNET are so rare that fewer than 50 cases have been reported till date in literature.

CASE HISTORY

A 38-year-old male presented with complaint of heaviness in left side of abdomen since 2 months associated with mild discomfort. There was no history of pain abdomen, hematuria, fever, lower urinary symptoms, anorexia, weight loss, cough, or bone pain. On examination, there was a large firm to hard abdominal mass with irregular surface extending from left subcostal margin to left iliac fossa and crossing the midline at the level of umbilicus. PET-CT scan was suggestive of a large heterogeneously enhancing solid cystic lesion of size 23×22×17 cm (SUV max 11.1), occupying almost the entire left kidney with tumor thrombus in left renal vein and infra hepatic inferior vena cava(IVC) (SUV max 9.1) (Figure 1). Minimal basal pleural effusion and pericardial effusion was noted and few sub centimeter aortocaval and paraaortic lymph nodes were also detected. Radical nephrectomy, lymphadenectomy and IVC thrombectomy was done. Patient received cyclophosphamide-based chemotherapy post operatively.

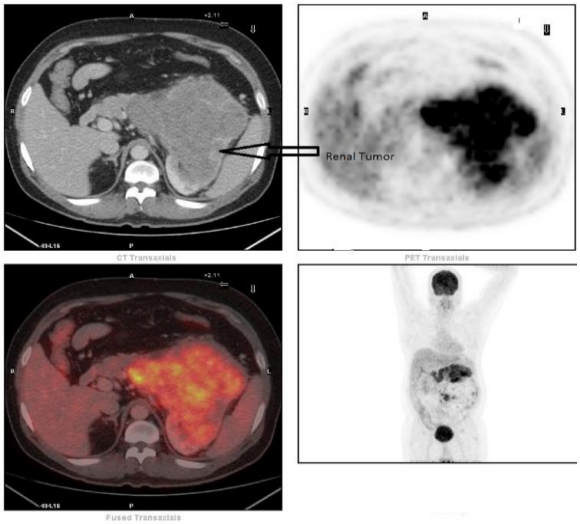

Figure 1: PETCT image showing tumor arising from left kidney

Ex-vivo the tumor mass measured 27cm in its largest dimension and weighed 4.6kgs.

Histopathology report was suggestive of PNET. Cells in IVC thrombus were also suggestive of PNET. Tumor involved renal sinus, renal vein, pelvis and perinephric fat. Gerota's fascia and ureteric cut margin was free of tumor. Tumor cells expressed CD99, Bcl-2, and Vimentin on Immunohistochemistry (Figure 2,3). They were negative for CD10, AMACR, CK (MNF), CK7, CD56, EMA and Synaptophysin. Chromosomal study was positive for EWSR 1 gene. The patient is on regular follow up for last one year and he is recurrence free at present.

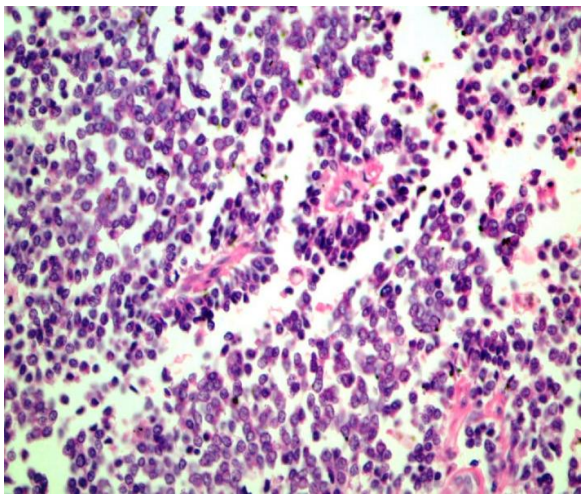

Figure 2: H and E stain showing spindle cells

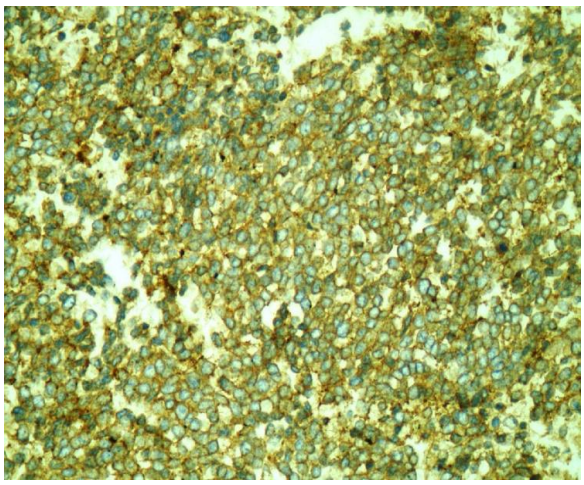

Figure 3: Immunohistochemistry showing CD99 positivity

DISCUSSION

PNET are a group of small round cell malignancies with neural crest origin. The incidence of PNET in abdomen and pelvis is about 14% of all peripheral PNETs. [1] Its origin is unknown. They may be part of neurocristopathies or may arise from primitive pluripotent stem cells. [1]

Primary renal PNET usually occurs at a median age of 27 year, and it has male dominance. [2] Age of our patient was 38 years, which is quite high as compared to the median age of presentation.

The clinical features of PNET may be non-specific like heaviness in abdomen, vague pain in flank or abdomen, weight loss, fever, lump in abdomen or hematuria. In our patient despite such a big mass, patient was relatively asymptomatic and just had complaint of abdominal discomfort.

Diagnosis and pathological staging is confirmed by histopathology, immunohistochemistry and cytogenetics in nephrectomy specimens.

As the differential diagnosis of PNET of kidney include other small round cell tumors like neuroblastoma, adult nephroblastoma, rhabdoid tumor, small cell sarcoma, round cell variant of synovial sarcoma and Wilms tumor, immunohistochemistry markers are used to confirm the diagnosis.

Histologically PNET is composed of small round cells which form Homer Wright rosettes or pseudo rosettes. Immuno histochemically these tumors are strongly positive for CD-99, Non-Specific Enolase (NSE) Vimentin, BCL-2, S-100, Synaptophysin. [1] Cytogenetic study play an important role to confirm the diagnosis, it shows balanced translocation t(11;22) (q24;q12) resulting in production of Ewing's Sarcoma Friend Leukemia Virus Integration 1 (EWS FLI-1)Fusion. [3]

On CT and MRI Renal PNET typically shows weak, heterogeneous enhancement, while clear cell and chromophobe RCCs shows moderate to marked enhancement. Multiple irregular enhancing septae are mostly seen in cases of PNET along with diffuse or peripheral hemorrhage and necrosis. [1] Calcifications are rarely seen in PNET.

On imaging, also can be seen tumor thrombus involving the renal vein and inferior vena cava, perinephric and retroperitoneal lymphadenopathy, liver and lung metastases, and bone metastases which appear to be lytic. [4] Involvement of adjacent organs may also be noted on imaging.

Aggressive Multimodality treatment regimen is recommended to manage these tumors. Radical nephrectomy with lymphadenectomy along with the removal of renal vein or inferior vena cava thrombus remains the mainstay of surgical management. Chemotherapeutic agents useful are vincristine, dactinomycin, Adriamycin, cyclophosphamide, ifosfamide, and etoposide. The current standard chemotherapeutic treatment of this group involves the use of a dose-intensive combination regimen that uses these 6 drugs in a modified protocol called Ewing's family of tumors (EFT)-2001. [2]

Radiation therapy may also play a role if complete surgical resection is not possible, presence of residual disease, positive margins or Gerota's fascia is involved. [5]

Despite aggressive treatment the prognosis of renal PNET is poor. The five-year disease-free survival rate in early disease is found to be around 45-50%, while in cases of advanced disease the median relapse period is around 1.5-2 years. [2]

Declaration of patient consent:

The authors certify that they have obtained all appropriate patient consent form. 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 his name and initials will not be published and due efforts will be taken to conceal their identity, but anonymity cannot be guaranteed.

Conflicts of interest: None
Financial support or sponsorship: None

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