

# Primary Ewing sarcoma/primitive neuroectodermal tumors of the kidney: Case series of eight cases from a single center with follow-up details

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## ABSTRACT

**Introduction:** We aim to share the experience of a single center in the management of eight cases of renal primitive neuroectodermal tumor (PNET) which are uncommon, aggressive tumors. The objectives were to study the presentation of the disease, the treatment offered and its outcomes, and the comparison of the treatment with published literature.

**Methods:** The single-center renal PNET data of all patients were retrospectively reviewed from 2011 to 2022. Renal PNET was seen in eight patients. Minimum follow-up period of 1 year was required.

**Results:** Male-to-female ratio was 7:1. The mean age was 26.5 years. All were locally advanced tumors on presentation. One patient had an inferior vena cava thrombus, one patient had metastases on presentation, and two patients had tumor extending to paranephric space. The diagnosis was made by histopathology supported by immunohistochemistry showing CD99 positivity. All patients were treated with radical nephrectomy, followed by chemotherapy in all and radiotherapy in three patients. Two patients expired at 3½ and 6 years after surgery, the remaining six are alive at a median follow-up period of 34.5 months.

**Conclusion:** Renal PNET is an uncommon renal tumor which is aggressive and requires multimodal therapy for prolonged survival.

## INTRODUCTION

Primitive neuroectodermal tumor (PNET) is characteristically a small round-cell malignant tumor derived from neuroectoderm. It is classified into central (CNS Central Nervous System - PNET) and peripheral Ewing sarcoma/PNET (ES/PNET). It has a signature alteration at the genetic level related to the EWS gene on chromosome 22q12. ES/PNET as a separate entity was described for the first time by Stout in 1918.<sup>[1]</sup> Ewing described the entity in detail in 1920.<sup>[2]</sup> ES/PNET is primarily a bone soft-tissue tumor. Primary renal PNET/ES is an uncommon tumor. A total of 362 cases have been reported in literature.<sup>[3]</sup> Seemayer was the first to write about renal PNET.<sup>[4]</sup> It presents at a young age,

is generally centrally located, aggressive, with early metastases resulting in poor prognosis, and requires multimodal therapy for treatment.<sup>[5-10]</sup> This report aims to describe experience in the management of eight cases of renal PNET.

## METHODS

Between 2011 and 2022, at a single center, 721 patients underwent radical nephrectomy for kidney tumor of whom, eight patients were diagnosed with PNET (1.1%). The study was done as per the Helsinki Declaration and its amendments following good clinical practice guidelines after taking the requisite approval from the institutional ethical

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committee (EC/742/2021). Written and informed consent for data sharing was taken from the participants included in the study. Patient demographics, clinical presentation, perioperative investigations such as blood reports and imaging in the form of contrast-enhanced computed tomography (CECT) scan of the chest, abdomen, and pelvis, and treatment received in the form of surgery, chemotherapy, with/without radiotherapy (RT), histopathological diagnosis, and survival on follow-up with follow-up imaging studies were noted. The authors confirm that all the original study data is available and accessible. Complications were graded as per the Clavien–Dindo system.<sup>[11]</sup> This case series has been reported as per the PROCESS 2020 guidelines.<sup>[12]</sup>

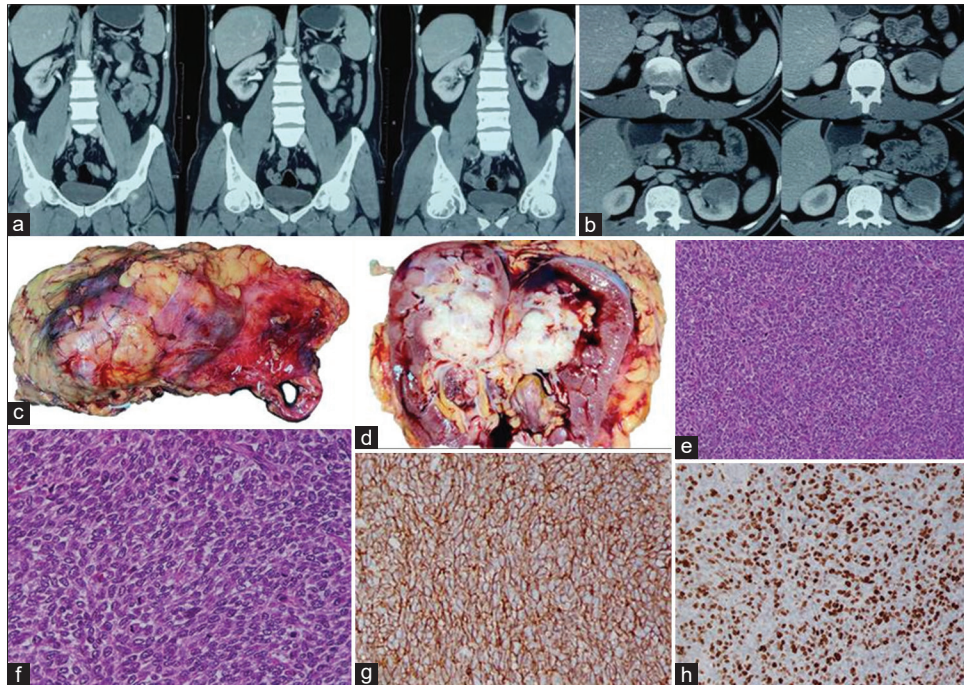
## RESULTS

Eight patients were included in the study Table 1. There was one female; on presentation, mean age was 26.5 years with a range of 4–46 years. All the cases had a tumor size >3.9 cm, one patient had tumor thrombus in the inferior vena cava (IVC), one patient had metastases on presentation, and two had tumors extending to paranephric space. On histopathological examination, all the cases showed small round blue cell neoplasm, and on immunohistochemistry (IHC), CD 99 staining was positive. Radical nephrectomy was done in all patients with laparoscopic approach in three patients and IVC thrombectomy was additionally performed in the patient with IVC tumor thrombus. In the postoperative

period, postoperative ileus of four or more days was observed in two patients, and blood transfusion was required in two patients resulting in Clavien–Dindo grade 2 complications, and the remaining patients had Clavien–Dindo grade 1 complications. Chemotherapy was required in all patients in the form of VAC/IE regimen (vincristine [V], adriamycin [A], cyclophosphamide [C], ifosfamide [I], and etoposide [E]) of total 16 courses for a duration of 1 year. One patient is receiving chemotherapy and the remaining all have completed their chemotherapy course. Three patients required local RT postoperatively, with two patients requiring it for residual disease and one patient needing it for recurrent disease. Posttreatment metastases were detected in one patient who expired after 3½ and the patient with preexisting metastases on presentation expired at 6 years after surgery due to malignant cachexia. The median follow-up duration was 34.5 months. The cancer-specific survival rate was 75% at 3 years and 75% at 5 years and using Kaplan–Meir estimates, cancer-specific average survival was 5.36 years and median survival is 6 years. CT scan images and biopsy specimen gross and histopathological slide images and IHC stain slide images are shown for three representative cases as can be shown in Figures 1-3.

## DISCUSSION

Primitive neuroectodermal tumors and Ewing sarcoma were initially considered separate entities but are now

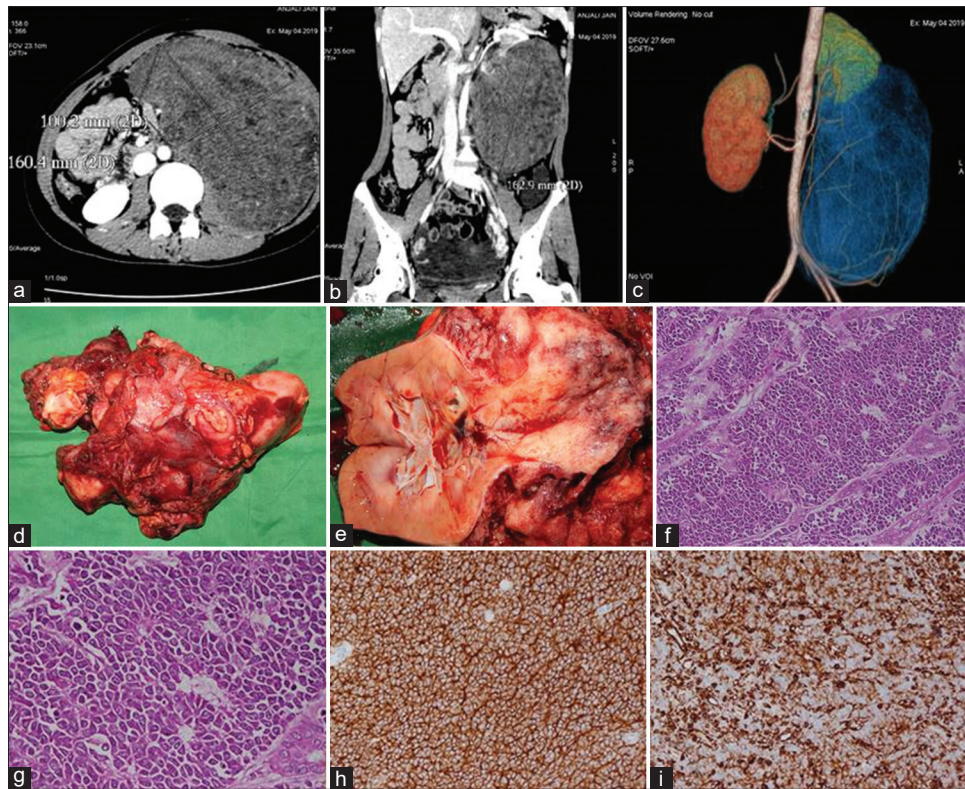


**Figure 1:** (a and b) contrast-enhanced computed tomography abdomen and pelvis coronal and axial views suggestive of a 6 cm × 4 cm × 4.3 cm × 4 cm well-circumscribed poorly enhancing renal mass at anterior cortex of left kidney with small cystic component and fine calcification extending to pelvic/cecal system and multiple enlarged left hilar nodes, largest 2.5 cm. (c) 15.5 cm × 9 cm × 7.5 cm size gross nephrectomy specimen weighing 426 g. (d) Gross specimen on bisection, left kidney, unifocal, infiltrating grayish white tumor in upper and middle pole, 5.5 cm × 4.5 cm × 4.0 cm, with solid and cystic areas with hemorrhage and necrosis are seen. (e and f) Low-power and high-power microscopy view of a section showing tumor composed of sheets of small round blue elongated cells with scant cytoplasm and round-to-oval pleomorphic nuclei with granular chromatin and thin vascular channels and scattered mitosis. (g) Immunohistochemistry (IHC)– CD 99 diffusely and strongly positive. (h) IHC– Ki 67 diffusely and strongly positive

**Table 1: Data of first eight patients**

Patient Details	Case 1	Case 2	Case 3	Case 4	Case 5	Case 6	Case 7	Case 8
Age (years)	42	17	35	39	46	7	22	4
Gender	Male	Female	Male	Male	Male	Male	Male	Male
Clinical presentation	Flank pain	Flank pain, abdominal swelling	Incidentally detected	Flank heaviness	Hematuria, weakness, loss of appetite	Flank pain, nausea, low-grade fever	Flank pain	Abdominal pain, hematuria
Clinical stage	T3a1T1Mx	T2bNx	T2bNx	T3aNxM1	T3bNx	T3aNx	T4N1	T4Nx
CECT scan findings	6 cm x 4 cm x 4.3 cm, well defined, poorly enhancing renal mass at anterior cortex of left kidney with small cystic component and calcification extending to pelvicalyceal system and multiple enlarged hilar nodes, largest 2.5 cm	16 cm x 10 cm x 16.2 cm well defined, poorly enhancing renal mass with exophytic component from lower pole of left kidney causing compression of surrounding structures	15 cm x 14 cm x 14 cm heterogeneously enhancing renal mass with necrotic areas in mid and lower pole of right kidney with exophytic component with compression of surrounding structures	10.5 cm x 7.1 cm x 6.9 cm heterogeneously enhancing mass in left kidney extending to renal hilum and perinephric space, hemorrhage in perinephric space, compression of surrounding structures, lung and liver lesions	9 cm x 6 cm x 6 cm, irregularly lobulated, mildly heterogeneously enhancing soft tissue mass in anterior and medial part of mid pole of kidney with extension to renal hilum and encasement of renal artery and vein	3.9 cm x 3.7 cm well defined, heterogeneously enhancing solid soft tissue mass in anterior and medial part of mid pole of kidney with extension to renal hilum and encasement of renal artery and vein	15 cm x 14 cm x 13 cm lesion, from medial cortex of mid and upper pole of left kidney with heterogenous enhancement and necrosis. Parapelvic extension. Pararenal and suprarenal space extension left anteriorly, encasing renal artery and splaying of renal vein and PCS 2 cm hilar lymph node	20 cm x 12 cm x 15 cm inhomogeneously enhancing mass from medial cortex of left kidney with mild HDN extending to pararenal space
IHC	CD 99 - positive, +3. Ki 67 - positive, +3	CD 99 - positive, +3. Vimentin - positive, +1	CD 99 - positive, +3. EMA and Vimentin - positive, +3	MIC2 - positive, +3 CD 99 - positive, +3	CD 99 and Vimentin - positive, +3	CD 99 - positive, Ki-67 - positive, S-100 - negative	CD 99 - positive +2, Ki 67 - positive, S-100 - negative	CD 99 - positive, +3, S-100 - negative
Surgery	Laparoscopic left radical nephrectomy	Left open radical nephrectomy	Right open radical nephrectomy	Preoperative angioembolization followed by open left radical nephrectomy	Open left radical nephrectomy and IVC thrombectomy	Laparoscopic right radical nephrectomy	Laparoscopic left radical nephrectomy	Open left radical nephrectomy
Postsurgery complications (Clavien-Dindo grade)	Grade 1 (nausea, vomiting)	Grade 1 (severe pain)	Grade 1 (paralytic ileus)	Grade 2 (blood transfusion)	Grade 1 (paralytic ileus)	Grade 1 (nausea, vomiting)	Grade 1 (severe pain)	Grade 2 (blood transfusion)
Chemotherapy	VAC/IE	VAC/IE	VAC/IE	VAC/IE	VAC/IE	VAC/IE	VAC/IE	VAC/IE
Radiotherapy	Yes, for residual disease	Yes, for residual disease	No	Yes, for local recurrence	No	No	No	No
Status on follow-up	Received post-RT chemotherapy IE, last CT scan was normal at 2 years of surgery	Alive at 4 years with normal CT scan on follow-up	Expired at 3.5 years, CKD status, developed metastases to lungs and liver, took alternative therapy after completing 1 year of chemotherapy	Patient expired at 6 years due to cancer cachexia	5-year follow-up, normal CECT	1-year follow-up, normal CECT	1-year follow-up, normal CECT	1-year follow-up, CECT scan awaited

CT = Computed tomography, CECT = Contrast-enhanced CT scan, PCS = Pelvicalyceal system, IVC = Inferior vena cava, HDN = Hydronephrosis, IHC = Immunohistochemistry, VAC = Vincristine, adriamycin, cyclophosphamide, IE = Ifosfamide and etoposide, RT = Radiotherapy, CKD = Chronic kidney disease, EMA = Epithelial Membrane Antigen, MIC: Microneme Protein



**Figure 2:** (a-c) Contrast-enhanced computed tomography scan of the abdomen showing axial, coronal and angiographic images showing a 16 cm × 10 cm × 16.2 cm well-defined, poorly enhancing renal mass with exophytic component from the lower pole of the left kidney causing compression of surrounding structures, not extending into pelvicalyceal system. (d) 13 cm × 10 cm × 8.5 cm left radical nephrectomy specimen weighing 412 g. (e) On bisection, reveals a left renal lower pole mass measuring 9 cm × 9 cm × 7 cm with hemorrhage is seen. (f and g) Low-power and high-power microscopy view of a section showing tumor containing Homer Wright rosette formation and sheets of small round blue elongated cells with scant cytoplasm and round-to-oval pleomorphic nuclei with granular chromatin and thin vascular channels and scattered mitosis. (h) Immunohistochemistry (IHC)– CD 99 diffusely and strongly positive. (i) IHC– Vimentin diffusely and strongly positive

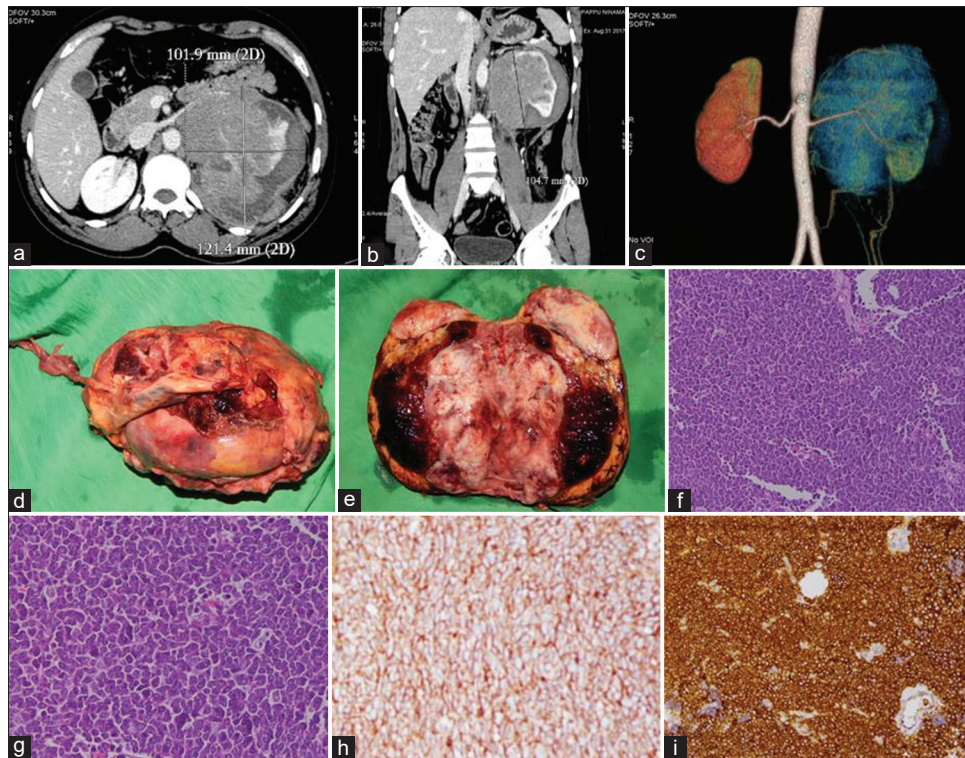
considered to be a spectrum of malignant conditions termed under the Ewing sarcoma family of tumors (ESFTs) due to better diagnosis by newer technologies such as cytogenetic and molecular studies.<sup>[3]</sup> ESFT includes various tumors such as Ewing sarcoma of bones, extraskeletal Ewing sarcoma, PNET, atypical Ewing sarcoma, Askin’s tumor, and peripheral neuroepithelioma.<sup>[13,14]</sup> Renal PNET constitutes 1.5% of all ESFTs as per Zöllner *et al.*<sup>[15]</sup> However, this could be an underestimation as there has been a steady increase in the incidence in recent literature probably due to better IHC-based pathological diagnosis.

In a recent meta-analysis of 356 patients by Bradford *et al.*,<sup>[3]</sup> the mean age at presentation was 28 years and the male-to-female ratio was 2:3 for patients with age <18 years and it was 3:2 in patients with age >18 years with 55% being males. In our study, the mean age at presentation is 26.5 years, and male-to-female ratio was 7:1.

In our study, the clinical presentation was left flank pain and swelling in six patients, an incidentally detected mass in one patient, and one patient had constitutional symptoms in the form of weakness, loss of appetite, and hematuria. Patients remain asymptomatic generally till the tumor reaches a large size, often reaching 10 cm in size and the

presenting symptoms then are flank pain, mass abdomen, hematuria, and constitutional symptoms.<sup>[16]</sup> The patients in our study presented with locally advanced disease with one patient having IVC thrombus, one patient having lymph node involvement, one patient with metastases, two patients with pararenal fat extension, and one patient developing distant metastasis after surgery. Against this, studies show that distant metastases on presentation is 53.2% population with the lungs being the most common site and lymph node involvement of 24% of patients on presentation, suggestive of aggressive behavior of this tumor.<sup>[3]</sup>

Diagnosing renal PNET preoperatively is difficult due to nonspecific presentation and radiological characteristics.<sup>[17]</sup> CECT scans of the abdomen in patients in our study were suggestive of large heterogeneously enhancing masses with cystic components, fine calcifications, hypo-dense necrotic areas, and hemorrhage within tumor. Some investigators have identified imaging features that are suggestive of PNET such as – large tumors, endophytic tumors that are infiltrative, multiple septae which are irregularly distributed, multiple areas of hemorrhage and necrosis, weak enhancement of tumor in relation to the renal cortex, and more chances of renal vein involvement and lymphatic and distant metastasis.<sup>[17]</sup> There is overlap



**Figure 3:** (a-c) Contrast-enhanced computed tomography scan of the abdomen showing axial, coronal and angiographic images show a 10.5 cm × 7.1 cm × 6.9 cm heterogeneously enhancing mass of left kidney extending to the renal hilum and perinephric space, hemorrhage in perinephric space, compression of surrounding structures, lung and liver lesions. (d) 16 cm × 11 cm × 10.5 cm left radical nephrectomy specimen weighing 831 g. (e) On bisection, reveals a 13 cm × 9 cm × 9 cm renal mass occupying the entire renal parenchyma with hemorrhage is seen. (f and g) Low-power and high-power microscopy view of a section showing tumor containing sheets of small round blue elongated cells with scant cytoplasm and round-to-oval pleomorphic nuclei with granular chromatin and thin vascular channels and scattered mitosis. (h) Immunohistochemistry (IHC)– CD 99 diffusely positive. (i) IHC– MIC (Microneme Protein) 2 diffusely and strongly positive

of these imaging findings with other kidney tumors such as renal cell carcinoma, nephroblastoma, neuroblastoma, lymphoma, and small round cell tumors (RCTs).<sup>[18]</sup>

Gross examination of the histopathological specimen reveals large, unilateral, poorly circumscribed tumor replacing renal parenchyma with a gray solid composition with areas of necrosis, hemorrhage, and cystic degeneration, locally extending to perinephric fat or renal vein.<sup>[13]</sup> In this study, we had similar nonspecific findings. On microscopic examination of the histopathological specimen, tumor cells are small round and blue with higher nuclear to cytoplasm ratio, round nuclei with barely visible nucleoli, and granular chromatin content. The morphological arrangement of cells is in the form of solid sheets, and lobules with finger-like infiltration into nearby normal renal tissue with Homer Wright rosettes.<sup>[13]</sup> These findings are also nonspecific and overlap with some renal tumors such as nephroblastoma, small cell tumors, malignant lymphoma, and some sarcomas such as clear cell and alveolar rhabdomyosarcoma.<sup>[19]</sup> IHC is mandatory for tumor confirmation. IHC profile of this tumor is like Ewing sarcoma in other locations. Diffuse membrane positivity to CD99, a macrophage inhibitory protein gene product is seen in >90% of patients. S100 and Vimentin are positive in 50%–70% of patients. Neuroendocrine markers such as neuron-specific enolase and synaptophysin positivity are seen in 48%–95%

of patients. Friend Leukemia Virus protein (FL1) positivity is present in 71%–84% of patients. Cytogenetics or PCR (Polymerase Chain Reaction) arrays or FISH (Fluorescent *In Situ* Hybridization) studies show EWS/FL1 fusion which helps to confirm diagnosis since it is seen in >90% of patients. It is due to chromosomal translocation from t(11:22) (q22:12). In our study, all patients were positive for CD99, and cytogenetic testing was not required for confirmation.

Standard therapy now is a multimodal therapy with radical nephrectomy with postoperative chemotherapy with/without adjuvant RT. Laparoscopic nephrectomy, which was first reported by Perer *et al.* in 2006,<sup>[20]</sup> is preferred wherever possible due to reduced patient morbidity and mortality. Chemotherapeutic agents used are vincristine (V), adriamycin (A), cyclophosphamide (C), ifosfamide (I), and etoposide (E). The current standard protocol is a dose-intensive combination protocol called Ewing's family of tumors (EFTs)-2001 for 1 year, which is modified from the earlier used RCT 2 protocol.<sup>[21]</sup> Adjuvant RT is given for positive resection margins, residual tumors, or recurrence.<sup>[22]</sup> Surgery is the most important part of multimodal therapy and before the use of chemotherapy, 5-year-survival was <10%.<sup>[23]</sup> In our study, three patients underwent laparoscopic radical nephrectomy, two patients received adjuvant RT for residual disease, and one patient received RT for recurrence, all

patients received chemotherapy. Newer treatment options are being studied for EFT, such as insulin-like growth factor antibodies to the receptor, RNA inhibition methods, CD-99 receptor antibodies, GSTM4 protein inhibition which is seen in the EWS/FL1 pathway and is identified in nonresponding patients to chemotherapy.<sup>[13]</sup>

Renal PNET prognosis remains poor because of nonspecific clinical and radiological presentation which makes a preoperative diagnosis difficult and the aggressive tumor biology with early local recurrence and distant metastasis to organs such as lungs, bone, and liver.<sup>[22]</sup> As per the meta-analysis by Bradford *et al.*, on presentation, there is metastasis in 53.2% of patients, lymph nodes are involved in 24% of patients, 1-year mortality is seen in 21.5% of cases, and 3-year mortality was 59.7%.<sup>[3]</sup> Even with aggressive treatment, the cure rate of such cancers is only 20%.<sup>[16]</sup> In our study, out of eight cases, one patient had metastasis on presentation, two patients have expired, two patients are undergoing chemotherapy postsurgery, and two patients are cured.<sup>[24]</sup>

Strengths of the present study were that the minimum follow-up duration was 1 year.

Limitations of our study were retrospective nature, single-center setting, small patient population, heterogeneity of treatment received, and short follow-up duration for three patients. Thus, definitive conclusions cannot be drawn.

## CONCLUSION

Renal ES/PNET is an uncommon neuroectodermal malignant soft-tissue tumor. Patients present at an advanced stage and have a poor prognosis due to aggressive tumor biology. Radiological features are nonspecific, and diagnosis can be confirmed only on histopathological examination with IHC. Treatment is multimodal therapy with surgery consisting of radical nephrectomy with chemotherapy with or without RT.

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