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Case Report

Primitive neuro-ectodermal tumour of kidney in adult: Report of four consecutive cases and review of the literature

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KEYWORDS

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Abstract *Background:* PNET of kidney is a rare entity and its diagnosis is complicated by the presence of a number of differential diagnoses. The disease is most commonly seen in young adults. Radical nephrectomy and adjuvant chemotherapy is the standard treatment. However, the patients have a modest survival and often develop distant metastasis. We herein report four cases of renal PNET (rPNET).
Methodology: We retrospectively retrieved treatment chart of four cases of rPNET.
Results: Median age was 29 years. Radical nephrectomy was performed in three cases. All four cases received multiagent chemotherapy. VAC alternating with IE was the commonest regimen. Compliance and tolerance to treatment was excellent. At the last follow up two patients were in complete remission whereas the remaining two cases had systemic metastasis and alive with disease.
Conclusion: Multimodality approach is required in rPNET. Patient with localized disease appears to have better disease control and survival.

Introduction

Primitive neuro-ectodermal tumours (PNET) are derived from the neural crest. These tumours mostly present as bone or soft tissue masses in the trunk or axial skeleton in children and young adults [1]. PNET of kidney (rPNET) is a rare entity and its diagnosis is complicated by the presence of a number of differential diagnoses like blastemal wilms, rhabdoid tumour, neuroblastoma, lymphoma and renal cell carcinoma. Because of rarity treatment approach has been extrapolated from the information of PNET of the trunk. Here, we intend to present four cases of rPNET treated by multi-modality approach.

Methodology

Patient information was retrospectively retrieved by reviewing hospital records from 2009 to 2014. Four patients of renal

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PNET were identified from the records. Information regarding the patient demography, treatment modalities and survival information was noted in predesigned proforma.

Results

Patient characteristics

The median age at presentation was 29 years (Range: 14–55 years). All four patients were male. The most common symptom was haematuria (75%) followed by abdominal pain (50%). Two patients presented with disease localised to the kidney. One patient had retroperitoneal lymphadenopathy while the other patient presented with suspicious lung nodule. All tumours were fairly large with median size of 8.2 cm × 6.5 cm (Range: 5.9–10 cm × 5.2–8 cm) (Fig. 1). Histological examinations revealed a small round cell tumour infiltrating renal parenchyma. The cells had scant cytoplasm, round nucleus with vesicular chromatin and conspicuous nucleoli. Tumour cells were immune positive for MIC2; and immune negative for Chromogranin in all four cases (Fig. 2).

Treatment

Surgery

Two patients with localised disease were treated by upfront radical nephrectomy with or without retroperitoneal lymph node sampling. The patient with small suspicious lung nodule also underwent radical nephrectomy with retroperitoneal lymph node sampling. So, 75% patients underwent a curative surgery. The details of patient characteristics and treatment outcome have been summarized in Table 1.

Chemotherapy

Adjuvant chemotherapy was prescribed for two cases and salvage chemotherapy was used in one case. The patient with retroperitoneal lymphadenopathy was treated with neoadjuvant chemotherapy. VAC alternating with IE was the commonest regimen used in 3 cases (75%). One patient was treated with ifosphamide and Etoposide.

Response to therapy and survival

Two patients had a complete response after primary therapy and had no evidence of disease at last follow up. The patient with retroperitoneal lymphadenopathy had a complete response to this treatment and was put on follow up. After six months, he developed a local recurrence which was treated with six cycles of ICE chemotherapy. He again had a complete response and was put on follow up. After three months, he developed multiple brain metastases and received whole brain radiotherapy. On last follow up, he had progressive disease. The patient with suspicious lung nodule at presentation was treated initially by radical nephrectomy followed by six cycles of adjuvant VAC alternating with IE regimen. He progressed on treatment and developed bone metastases for which he received palliative radiation. The median survival was 10.5 months.

Follow up

All patients were followed up at regular interval. For the first two years patients were seen every three months and subsequently every six months for up to five years. A CECT of the abdomen and X-ray of the chest were done in each visit in addition to clinical examination.

Pattern of recurrence and salvage therapy

Two patients progressed and developed systemic metastasis. One patient had brain metastasis while the other had lung and bone metastasis. Salvage chemotherapy was prescribed with ICE regimen for one patient with local relapse and achieved CR. However, he developed brain metastasis thereafter. The other patient developed lung and bone metastasis and was advised for best supportive care.

Discussion

Renal PNET (rPNET) is a rare tumour with less than 125 cases reported since 1975. It occurs in children and young adults with a male preponderance [1]. The tumours tend to be very large, often more than 10 cm in size [2–4]. Renal PNET behave more aggressively than PNET arising from other sites and 20–50% is metastatic at presentation [1,3]. The five year disease free survival has been reported to be 45–55% [5]. In our case series, the median age was 29 years and all were males. The median survival was 10.5 months.

The presenting symptoms are nonspecific and similar to other renal tumours. The imaging characteristics are also

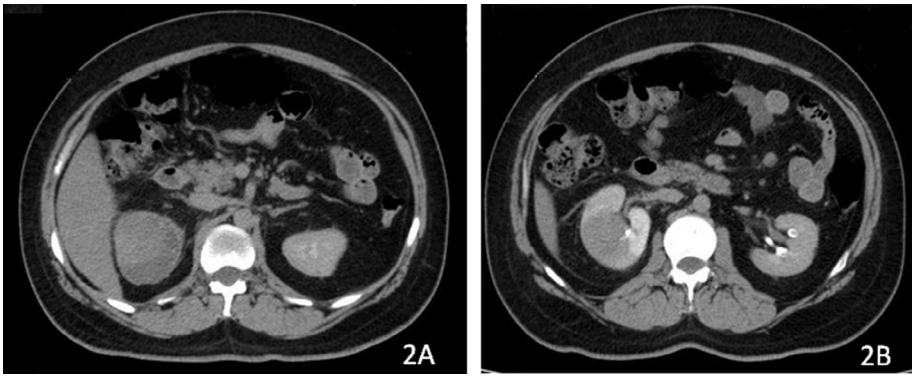

Figure 1 (A and B) Axial CECT scan image showing large contrast enhancing mass arising from the upper pole of the right kidney abutting the inferior margin of liver.