Primary Ewing's sarcoma of the kidney: A diagnostic and therapeutic challenge

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ABSTRACT

Ewing's sarcoma is a highly malignant tumor that usually arises from the bone and soft tissue of the extremity. Primary Ewing's sarcoma of the kidney is extremely rare. Here, we present the case of primary Ewing's sarcoma of the kidney in a 30-year-old man, presented with the left flank pain, and treated with the left radical nephrectomy with adjuvant chemotherapy. Despite the treatment options, the overall survival of the disease is poor. The relevant literature of this rare entity was reviewed.

Key words: *Ewing's sarcoma, Malignant renal tumor, Nephrectomy*

Ewing's sarcoma is a highly malignant tumor that usually arises from the bone and soft tissue of the extremity. Primary renal Ewing's sarcoma is extremely rare. Less than 100 cases have been reported in the literature [1,2]. It originates from neural and neural crest cells [1,3]. Historically, Ewing's sarcoma and primitive peripheral neuroectodermal tumor (PNET) were described as two different pathologic entities. Ewing's sarcoma and PNET are now considered as the same tumor; both have indistinguishable chromosomal translocation and immunohistochemical characteristics [4]. On the basis of imaging techniques, it cannot be differentiated from the renal cell carcinoma. Definitive diagnosis requires histopathology, immunohistochemistry (IHC), and cytogenetic study. Despite the treatment options, the overall survival is poor. Here, we present the case of a 30-year-old male with primary Ewing's sarcoma of the kidney which is an extremely rare entity.

CASE REPORT

A 30-year-old male presented with a complaint of the left flank pain for the past 1 month. The pain was dull aching, intermittent, mild in severity, and not associated with nausea and vomiting. He had no history of gross hematuria. His medical history was unremarkable.

On general examination, the patient was conscious, oriented, and fairly nourished. There were no signs of pallor, icterus, clubbing, cyanosis, edema, or lymphadenopathy. His vitals were within normal limits. On physical examination, there was a mass 15 × 8 cm occupying the left hypochondriac, lumbar, and the left iliac fossa region. The mass was moving with respiration, bimanually palpable, and ballotable.

All the blood investigations (hemoglobin – 14 gm%, total leukocyte count – 6600/mm^3, serum creatinine – 0.64 mg/dl, total bilirubin – 0.47 mg/dl, alkaline phosphate – 81 u/l, aspartate aminotransferase – 36 U/l, alanine aminotransferase – 43 U/l, lactate dehydrogenase – 236, and serum calcium – 8.7 mg/dl) and urine analysis were normal. The chest X-ray was unremarkable. Contrast-enhanced computerized tomogram revealed large exophytic soft tissue mass in the left kidney of size 8.4 cm × 11.7 cm × 14.6 cm showing heterogeneous contrast enhancement with the necrotic area (Fig. 1). There was no evidence of metastasis. He underwent the left radical nephrectomy with an uneventful post-operative recovery.

On cut section, the whole kidney was replaced by white mass and necrotic areas. Histopathologically, the tumor composed of diffuse sheets of round to oval cells with scanty cytoplasm and hyperchromatic uniform nuclei separated by fibrous septa (Fig. 2). Capsule, fascia of Gerota, and perinephric fat were involved by the tumor. IHC revealed the tumor cells positive for CD99 and negative for LCA, EMA, BCL2, and CD20 (Fig. 3). On the basis of histopathological examination and IHC, a diagnosis of Ewing's sarcoma of the kidney was made.

DISCUSSION

Ewing's sarcoma of the kidney is an extremely rare and highly aggressive tumor. It was first reported in 1975 by Seemayer *et al.* [3]. The usual age of diagnosis is between 28 and 34 years and it is more common in males [5,6]. Our patient was a 30-year-old male. Usually, these patients present with non-specific symptoms including flank pain, hematuria, and symptoms related to urinary tract infection [7-9], but in our case, the patient presented with the left flank pain only. As per Risi *et al*., 66% of patients present with metastasis at the time of diagnosis [7]. Lungs are the most common site of metastasis followed by liver and bone [10,11]. In the present case, there was no metastasis at the time of diagnosis.

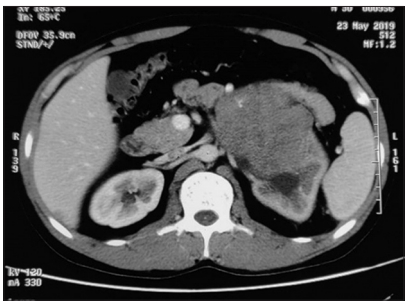

Figure 1: Contrast-enhanced computerized tomogram showing the left kidney mass with heterogeneous contrast enhancement and necrosis

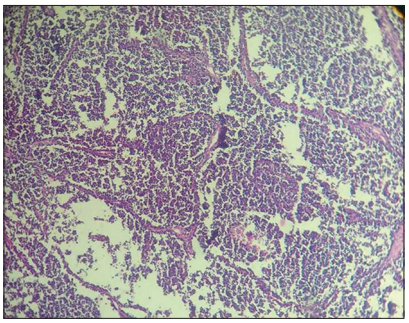

Figure 2: Histopathological examination shows that the tumor composed of diffuse sheets of round to oval cells with scanty cytoplasm and hyperchromatic uniform nuclei separated by fibrous septa H and E (×40)

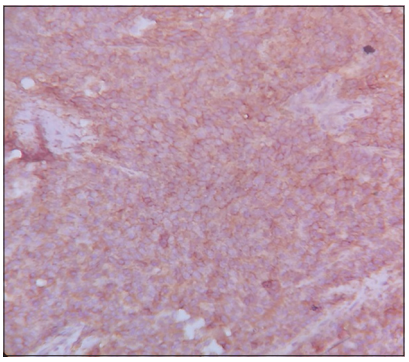

Figure 3: Immunohistochemistry revealed CD 99 positivity in tumor cells

Diagnosis is based on histopathology, IHC, and cytogenetic study. Histopathologically, Ewing's sarcoma is characterized by small round blue cells that involve necrotic areas. Expression of cell surface antigen CD 99 is present in 95% of cases of Ewing's sarcoma [12]. In the present case, CD 99 positivity was present. EWS/FLI-1 gene fusion resulting from translocation t (11:22) (q24;q12) is the gold standard for the diagnosis [1]. The differential diagnosis of small round blue cell tumor includes Wilms' tumor, neuroblastoma, and lymphoma, clear cell sarcoma of the kidney, and Ewing's sarcoma [13].

The treatment strategies include surgical resection, adjuvant chemotherapy, and radiotherapy. The effective chemotherapeutic agents include vincristine, doxorubicin, etoposide, ifosfamide, cyclophosphamide, and actinomycin D. In non-metastatic disease, the addition of etoposide and ifosfamide to doxorubicin-containing regimens has shown survival advantage [14]. Pre-operative diagnosis of Ewing's sarcoma of the kidney is difficult; therefore, radical nephrectomy was initial treatment in the majority of cases. Five-year disease-free survival rate was found to be 45–55% for tumor localized to the kidney, whereas the median relapse-free survival of patients with metastasis was only 2 years [2].

CONCLUSION

Ewing's sarcoma of the kidney is an extremely rare and highly malignant tumor. Definitive diagnosis requires cytogenetic study and IHC. Despite treatment, the overall survival is poor.

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