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Primary Ewings Sarcoma of Kidney: A Rare Tumor of Kidney

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Abstract
Ewings sarcoma/primitive neuroectodermal tumor represents a spectrum of undifferentiated tumors with similar biology that together represent the second most common sarcoma in the pediatric -young adult age group. Ewings sarcomas of the kidney are rarely found high-grade malignant tumors and very few cases have been reported to date. A range of aggressive small blue round cell tumors have been subsumed under the term Ewings sarcoma family tumor due to their shared undifferentiated round cell phenotype. . Establishing a correct diagnosis is critical because renal Ewing sarcoma carries a dismal prognosis and thus dictates a specific treatment strategy. We present a case of an adolescent boy presenting to us with flank pain. On evaluation, he was found to have right renal mass which was reaching up to liver parenchyma. A nephrectomy was done and final pathology report was Ewings sarcoma
Key words: *Ewings sarcoma; primitive neuroectodermal tumor;Renal ewings sarcoma*

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I. Introduction

Ewings sarcoma mostly affects adolescents and young adults. It is a genetic disease resulting from a translocation mutation fusing the EWS and FLI1 genes of chromosome 22 and 11, respectively. Ewings sarcoma arises most commonly in bone. Ewing sarcoma of the kidney is a rare tumor of the primitive neuroectodermal tumor family[PNET][1]. The presence of small round cells with rosettoid arrangement on pathology and CD99 and FLI-1 expression is critical evidences for diagnosis.

II. Case History

A 16-year-old male patient presented with a history of abdominal pain in the right lumbar region for the past 2 weeks. On clinical examination, a mass was palpable in the right lumbar region extending to the right hypochondrium. He was subjected to a computerized tomography[CECT] scan of the abdomen which showed a solid lesion with necrotic areas arising from the upper and mid pole of the kidney measuring 12.7*13.4*17.3 cm and the right kidney is near completely replaced by a mass lesion[FIG1]. An ultrasound-guided biopsy was done which came as ewings sarcoma. Positron electron tomography[PET]scan was done which showed no distant metastasis. Right radical nephrectomy was done. On gross examination, the kidney was replaced by hemorrhagic, necrotic material with cystic spaces and a small portion of residual renal parenchyma at the periphery[FIG 2]. Histologically tumor was comprised of small round cells arranged in sheets, occasional rosettes and large areas of hemorrhage and necrosis[FIG 3]. Immunohistochemistry[IHC] was done to rule out other renal tumors. Tumor cells show strong diffuse membranous positivity for CD 99 and FLI-1[FIG 4] and were negative for WT1 which was consistent with the diagnosis of ewings tumor

Patient was clinically stable after surgery and discharged on postoperative day 10 after removal of sutures and abdominal drain .Pateint was referred to medical oncology department for adjuvant chemotherapy

III. Discussion

Renal sarcomas are rare malignancies that constitute less than 1% of all malignant renal tumors. Sarcomas of the kidney usually remain asymptomatic until they are large enough to produce symptoms, the average size at time of diagnosis varies from 5.5cm to 23 cm

The majority of patients affected by EWS of kidney are in their second and third decade of life with slight male predominance[2]. Presenting complaints and radiological imaging is non-specific and can mimic other renal mass lesions such as Wilms tumor and Renal cell carcinoma[3]. . This tumors appear ill-defined,large heterogenous masses with necrotic and hemorrhagic areas

The clinical findings are uncharacteristic and patients usually complain about pain, palpable masses, and hematuria. Grossly renal EWS presents with areas of necrosis and hemorrhage along with cystic degeneration[4].In our case, we had similar gross findings.

The principal management for renal EWS is surgical resection followed by adjuvant chemotherapy. This management was reinforced by a case reported by Ohgaki et al of a 21-year-old man who underwent only surgical resection for renal EWS and later got systemic recurrence. Our patient was also managed by upfront surgery and adjuvant chemotherapy. Chemotherapeutic agent similar to that used for osseous EWS such as doxorubicin, vincristine, and cyclophosphamide alternating with ifosfamide and etoposide is the current standard chemotherapy[5].

Malignant round cell tumors of the kidney can be a diagnostic challenge, particularly in children. The differential diagnosis can include Wilms tumor, Neuroblastoma, Ewings sarcoma, poorly differentiated synovial sarcoma, clear cell sarcoma of the kidney, and, Non-Hodkins lymphoma[6]. The entrapped tubules can undergo cystic change and can also have an embryonal look, leading to the possibility of Wilms tumor. In this case monomorphic population of cells, occasional rosettes, abundant necrosis with peripheral sparing of tumor cells were the clues for diagnosis.IHC is used to confirm the diagnosis. In our case tumor cells were positive for CD 99, FLI-1 and negative for WT1 and TLE1.PNET cells can also show NSE positivity but neuroblastoma cells are always CD99 negative.CD 99 is also positive in synovial sarcoma but it will also show positivity for BCL-2 and CK which was negative in our case. Approximately 85-90% of EWS can be defined by a DNA translocation t(11;22)(q24;q12)[7]. The resultant gene fusion creates a functional oncogene. Histologically EWS and small cell carcinomas can form Homer -Wright rosettes. The gene fusion EWS/FLI1 can be used to distinguish between these two entities[8].

IV. Conclusion

Primary EWS of the kidney is a rare diagnosis. Renal PNET occurs predominantly in young adults and tends to be extremely aggressive. Diagnosis is based on the histologic, immunohistochemistry and molecular findings. Treatment strategies includes surgery, chemotherapy, and radiotherapy. Based on our case and review of the literature we recommend upfront surgical resection with chemotherapy. Post-op radiotherapy may be added if regional lymph nodes are enlarged. Patients without metastasis have shown excellent survival. The prognosis of patients with metastases is poor with overall survival rate of 5.6 months, hence early detection and multimodal treatment are indicated.

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Caption for Images

FIG1.CECT scan showing right renal mass abutting and displacing right lobe of liver

FIG 2.Gross specimen of right kidney showing haemorragic and necrotic material with cystic spaces.

FIG 3.Histopathology of tumor showing monomorphic population of small round cells arranged in sheets with ocassional rosettes.

FIG 4.Immunohistochemistry of tumor showing positivity to CD99 and FLI-1

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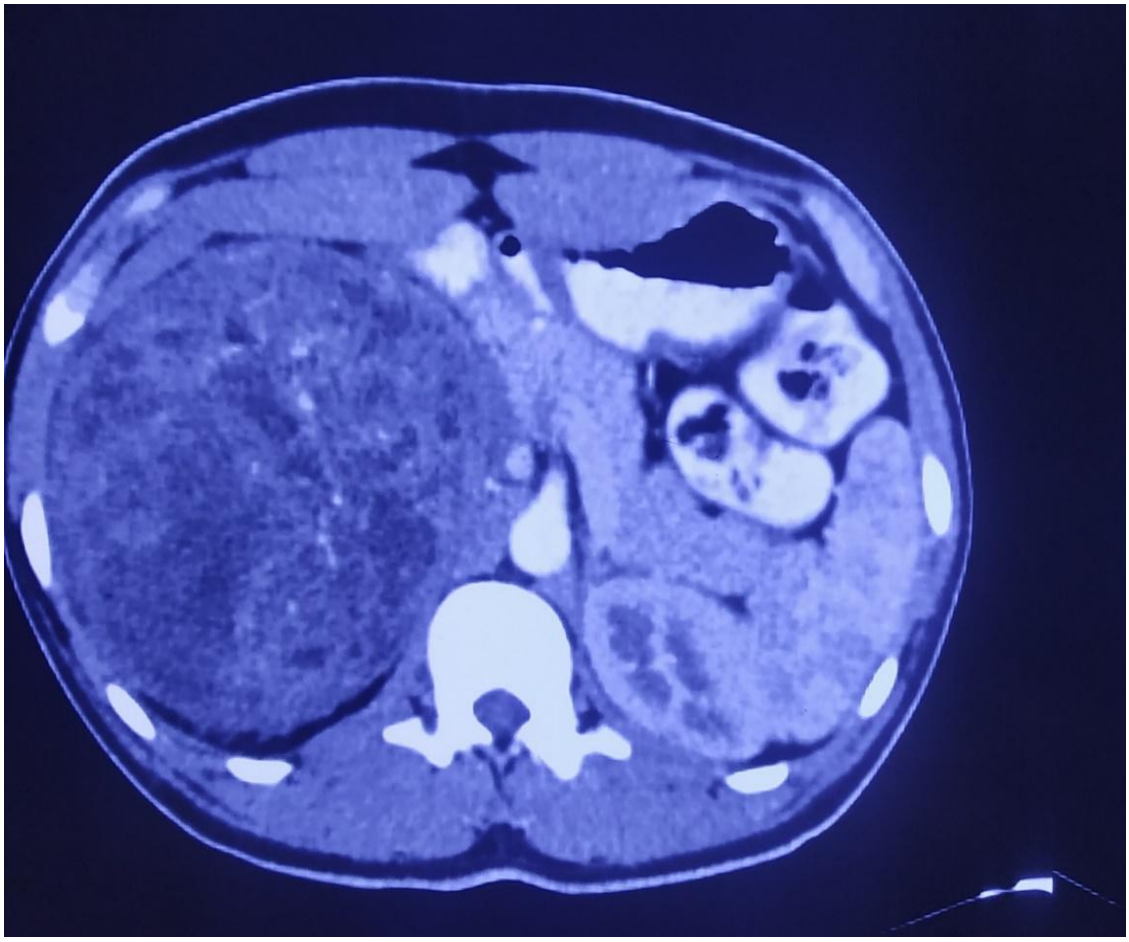


FIG 1

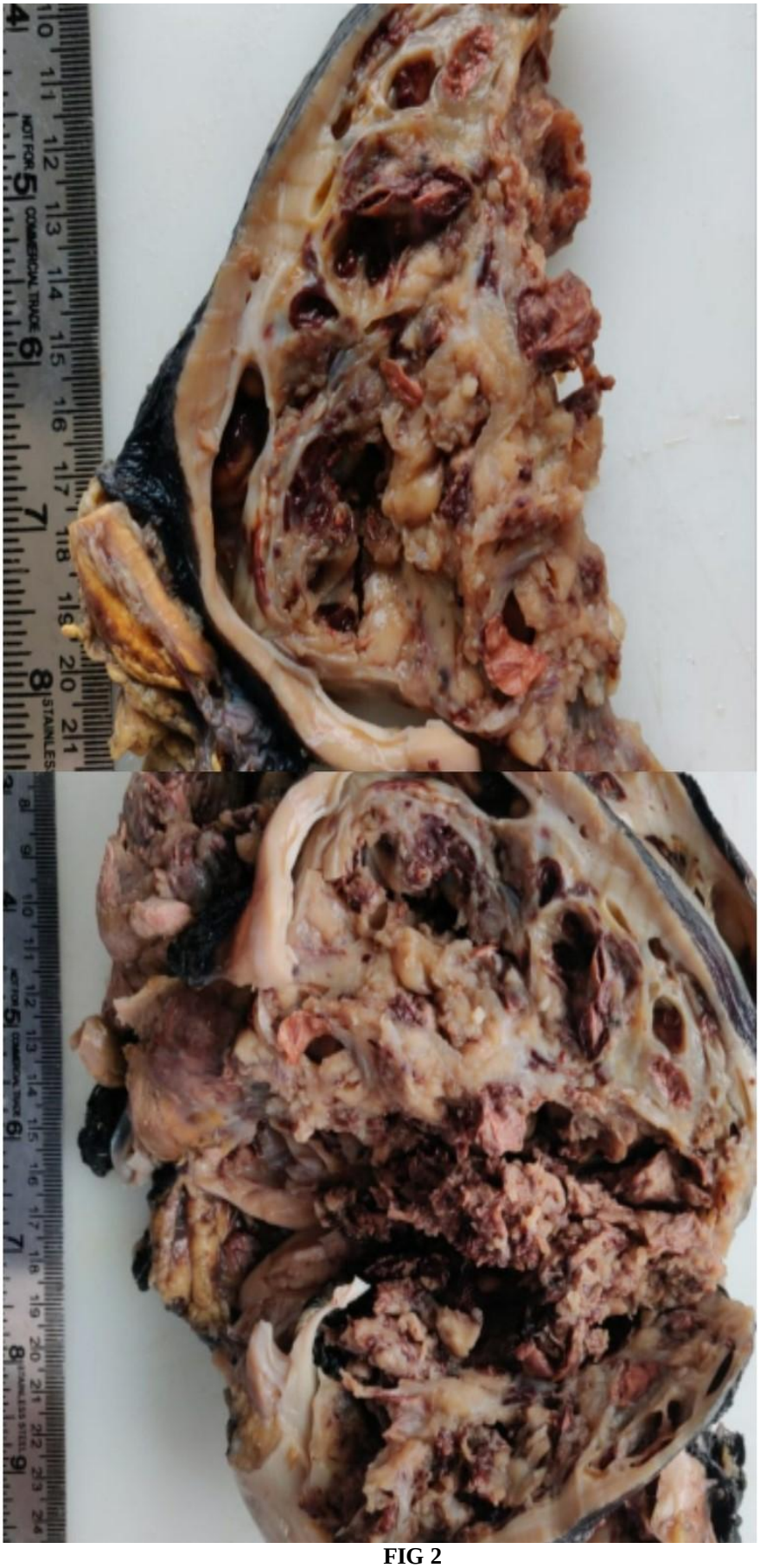


FIG 2

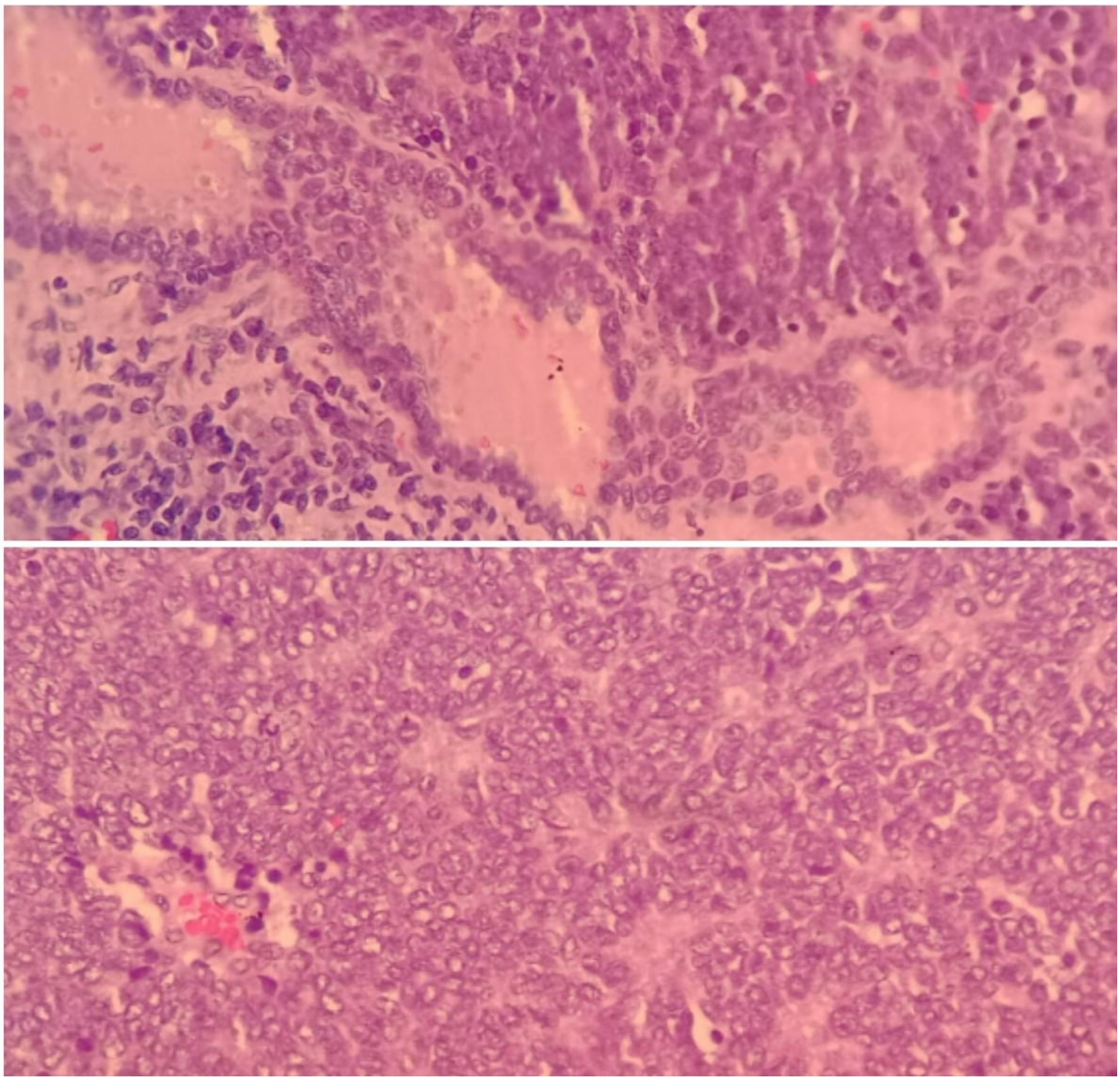
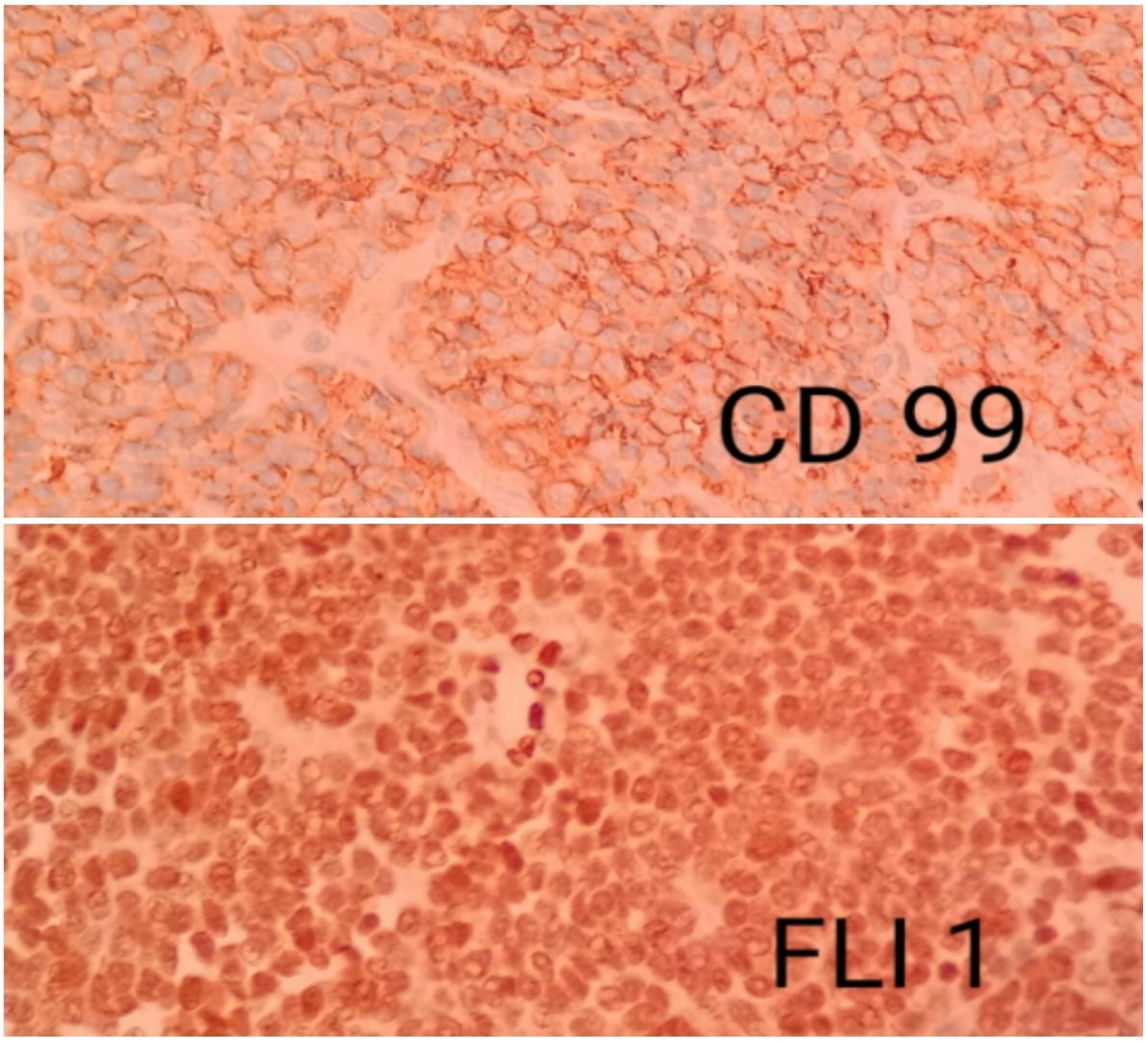


FIG 3



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