Case Report
ISSN 2521-8492

Ewing Sarcoma of the Kidney; a Rare Case Report

Anmar Alharganee[1*], Haydar Alabidi[2], Sarah Alkubaisi[3]

[1]Department of Oncology, Nottingham University Hospital, Nottingham, UK.
[2]Oncology Teaching Hospital, Baghdad Medical City; Radiation and Nuclear Medicine Hospital, College of Medicine Baghdad University, Baghdad, Iraq.
[3]Diagnostic Radiology Specialist Jordan University Hospital, Amman, Jordan.
*Correspondence to: Anmar Alharganee (E-mail: dr_harganee@yahoo.com)
(Submitted: 05 June 2020 – Revised version received: 24 June 2020 – Accepted: 19 July 2020 – Published online: 26 September 2020)

Abstract
Kidney cancer is considered as one of the commonest urological malignancies globally. Primitive neuroectodermal tumor and ewing sarcoma pathology are commonly found bone cancers within the daily clinical practice especially in adulthood. In this article we will discuss a rare case report of ewing sarcoma family variant (ESFV) within the renal pelvis. Chemotherapeutic and radiation protocols already used in bone ewing sarcoma have been exploited to treat this variant pathological malignancy in the kidney considering the metastatic presentation of the case. Hence, the response to some of these management lines was obvious during follow up appointments. Unfortunately, side effects were so severe especially the hematological adverse events. By conclusion, the administration of the same protocols as for bone ewnig sarcoma should be discussed carefully in treatment selection.
Keywords: ESFV, neuroectodermal tumor, ewing sarcoma

Introduction

Primitive neuroectodermal tumour or ewing sarcoma family variant are mainly described as an inseparable pathologically. However, their unique stem cell progenitor cells and chromosomal alterations made them known as the Ewing Sarcoma Family Tumor or ESFT.[1]

The presence of ESFT in the kidney is very unusual and this include neural cells that invigilating the embryological development of the kidney.[2]
Generally, sarcomas in the kidney are considered extremely rare mainly leiomyosarcomas and liposarcomas with figures at 60% and 15%, respectively.[3]

We will present a case of a 37 years old female diagnosed with advanced neuroectodermal tumor of the kidney and the adrenal gland.

Case Report

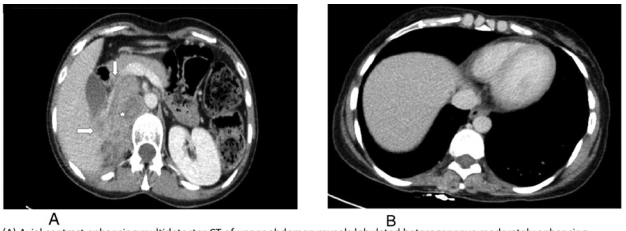
Patient is a 37 years old female with no past medical history presented with sudden onset sharp right sided flank pain and shortness of breath. This was clinically evaluated and by imaging ultrasound that revealed an upper pole mass in the right kidney and right sided pleural effusion. These findings were confirmed by CT scans of the chest and abdomen that showed an ill-defined heterogeneous mass about 9.6 *8.5 cm in the renal area.

After biopsy, pathology has shown sheets of small round primitive cells with extensive areas of necrosis. Besides, Immunohistochemistry revealed Areas from the sample strongly and diffusely positive for CD 99 and NKX2. While, negative for WT, CK, CK7, S-100 and Desmin. Diagnostic pleural fluid aspiration showed diffuse malignant cells consistent with the primary finding. After discussing treatment options with the patient she was started on palliative chemotherapy VAC protocol (Vincristin, Doxorubicin and Cychlophosphmide) for six cycles every 21 days. After 2 cycles of the aforementioned protocol patient reported no shortness of breath, weight gain, no fatigue and increase in appetites. After completing the whole 6 cycles there was a regression of the renal mass and complete resolution of the malignant pleural effusion.

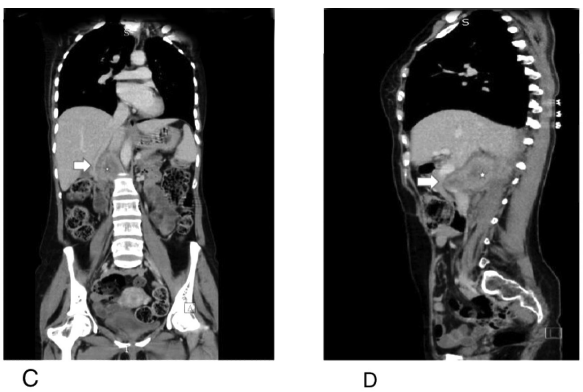
Patient received radiation to the kidney mass and then kept on follow up.

After 4 months of regular follow up and stable disease the patient were progressing symptomatically, clinically and by imaging.

The decision was to start second line IE protocol (Ifosfmide and Etoposide) which was intolerable by the patient after the second cycle of therapy due to severe hematological toxicity and life threatening febrile neutropenia that need frequent hospitalizations. Patient opted to keep on

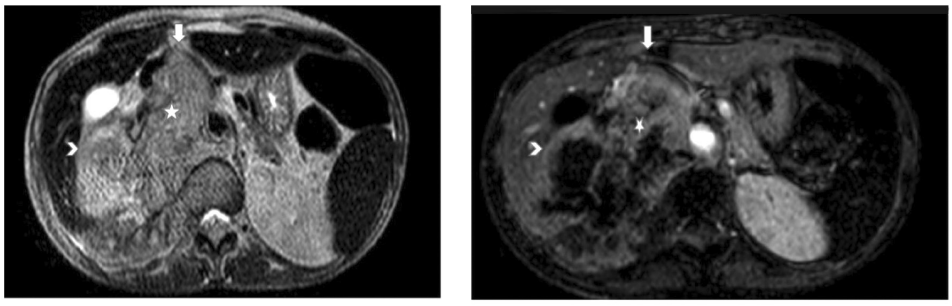


(A) Axial contrast enhancing multidetector CT of upper abdomen reveals lobulated heterogeneous moderately enhancing mass at the anatomic site of right kidney (star) with necrotic areas obliterating the hepatorenal space exerting mass effect and stretching the right renal artery displacing it anteriorly (vertical arrow) , no definite hepatic invasion , this mass now measures 6.5 cm CC * 4.5 cm trans.* 8 cm AP (has regressed in size compared to CT done 6 months ago) , A metastatic necrotic lymph node seen within the portahepatis (Horizontal arrow)
(B) Disappearance of the malignant pleural effusion



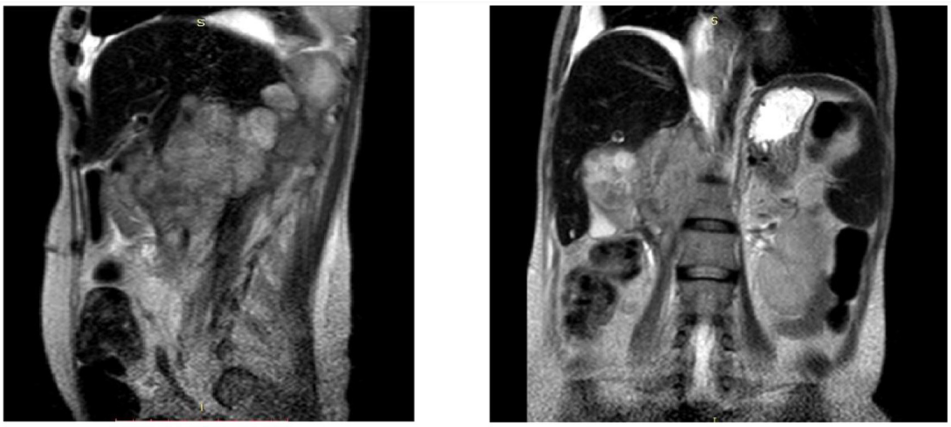
(C) Coronal (D) sagittal reconstructed CEMDCT reveals the same moderately enhancing mass with necrotic center (star) compressing and narrowing the IVC (horizontal arrow) , with clear cleavage from the liver , No pleural effusion

Fig. 1



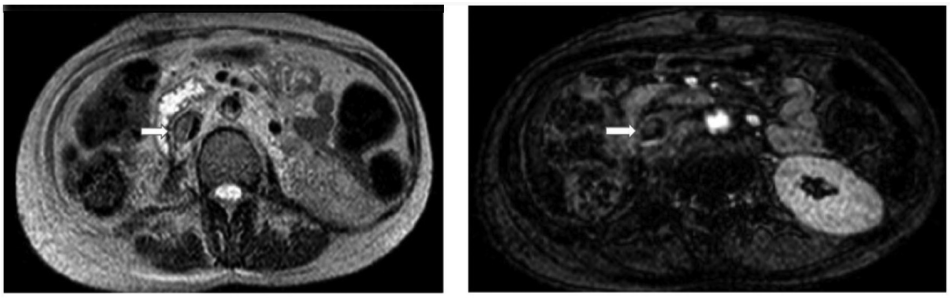
A B

(A) Axial T2WI with fat suppression done 3 months later reveals lobulated heterogeneous isointense mass at the anatomic site of right kidney appearing larger compared to previous with internal necrotic areas (star) , further anterior displacement of the right renal artery (vertical arrow) , no hepatic infiltration , portahepatis lymph node appearing larger (arrow head) , with loss of signal void of IVC (B) T1WI with IV Gadolinium contrast heterogeneous enhancement of the mass with necrotic centers



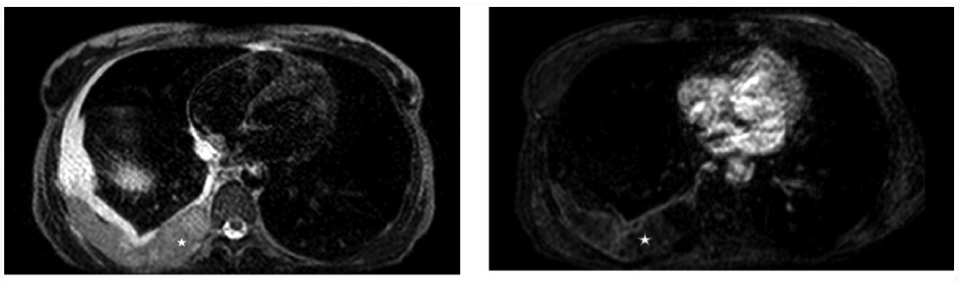
C D

(C) ,(D) MRI T2WI coronal, sagittal reformations



E F

(E) Axial T2WI (F) Axial T1WI with Gadolinium IV contrast of the mid abdomen reveals newly developing partial filling of the IVC (arrow) with an isointense eccentric thrombus that showed moderate enhancement following contrast injection in keeping with tumoral thrombosis



G H

(G) Axial T2WI Fat Sat. (H) Axial T1WI with IV Gadolinium contrast of the lower chest reveals newly appearing thick nodular solid pleural masses of posterior segment of right lower lobe showing moderate enhancement following contrast injection (star) in keeping with pleural metastasis , development of mild right sided pleural effusion

Fig. 2

continuing the treatment chemotherapy protocol and asked for other alternatives. She was started then on the third line TC protocol (Topotecan and Cychlophosphmide) regimen. Patient gradually responded very well symptomatically in terms of improvements in complete blood count, increase in weight and advanced physical activity and kept on regular follow up.

Discussion

PNET/EWST of the kidney are unfamiliar urological malignancies and occurred predominantly in early adult life.[4] Chemotherapeutic agents including Anthracyclin based protocols may reveal a response in these kind of tumors and mainly develop symptomatic resolution of the disease. By contrast, the protocols usually used to treat ewing sarcoma within the bone when applied in similar cases in the kidney are considered to be more toxic to the patient and mainly cause life threatening hematological side effects.

Conclusion

Cases of visceral PNET/EWST are difficult to be diagnosed pathologically and required an experienced physician to confirm the pathology especially in the kidney using different panels of immunohistochemistry markers.

Management decisions should be supported on and discussed in a multidisciplinary meeting and carefully explained to patients especially in terms of prognosis and treatment related adverse effects. In addition, recruiting patients in clinical trials to explore a targeted therapy is highly recommended to achieve treatment personalization.

Thus, treatment side effects should be highly considered when discussing therapy options in visceral EWST and when guiding therapeutic possibilities.

Conflicts of Interest

None.

References

1. Ellinger J, Bastian PJ, Hauser S, Biermann K, and Muller SC. "Primitive neuroectodermal tumor: rare, highly aggressive differential diagnosis in urological malignancies". Urology, Vol. 68, no2, PP. 257–262, 2006.
2. Parham D, Roloson G, Feely M, Green D, Bridge J, Beckworth J, (2001). Primary malignant neuroepithilia tumor of the kidney: a clinic-pathological analysis of 146 adult and pediatric cases from the National Wilms: Tumor Study Group Pathology Center. Am J Surg Pathol 25:133–146.
3. Lalwani N, Prasad SR, Vikram R, KatabathinqV, Shanbhogue A, and Restrepo C. "Pediatric and adult Primary sarcoma of the kidney: a cross sectional imaging review. Acta Rdiologica, vol. 52, 4, pp. 448–457, 2011.
4. Bing Z, Zhang P, Tomaszewski J, Maclennan G, (2009). Primary Ewing Sarcoma primitive neuroectodermal tumor of the kidney. J Urol 181: 1341–1342.