ORIGINAL RESEARCH PAPER
Volume - 12 | Issue - 11 | November - 2023 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

RENAL EWINGS SARCOMA/PRIMITIVE NEUROECTODERMAL TUMOR: A RARE CASE REPORT AND REVIEW OF LITERATURE

Histopathology

Mitunkumar Mayani Senior Resident, Deparment Of Pathology, Santokba Durlabhji Memorial Hospital, Jaipur, Rajasthan.

Smitirupa Mishra Resident, Deparment Of Pathology, Santokba Durlabhji Memorial Hospital, Jaipur, Rajasthan.

Rohit Jain Senior Consultant, Deparment Of Pathology, Santokba Durlabhji Memorial Hospital, Jaipur, Rajasthan.

ABSTRACT

Primitive neuroectodermal tumor (PNET) is a malignant small round cell tumor which typically arises from bone or soft tissue in adolescents and young adults, and is equivalent to Ewing's sarcoma. Renal PNET is extraordinarily rare and exhibits highly aggressive biological behavior with poor prognosis. The clinical presentation and imaging findings are non specific and are not characteristic. Very few cases have been reported in literature hence we present a case of 21 years old male whose radical nephrectomy specimen was received in our department for histopathological examination. The histomorphological characteristics and immunohistochemical analysis confirmed the diagnosis of renal PNET.

KEYWORDS

INTRODUCTION

Primitive neuroectodermal tumor(PNET) is a malignant small round cell tumor and typically arises from bone or soft tissue in the trunk or axial skeleton in adolescents and young adults. Localization of PNET in genitourinary system such as kidney, bladder and prostate is rare[1].

As a member of the Ewing's sarcoma family of tumors (ESFTs), primitive neuroectodermal tumors are rare kind of sarcoma, especially those found as primary tumors in kidney are exceedingly rare and fewer than 200 cases are reported in medical literature so far[2].

Primary Renal PNET has highly aggressive clinical course than other sites with high propensity for early metastases and death[3]. Differential diagnosis includes the other members of small round cell "blue tumor" family which can impose diagnostic difficulties due to frequent overlapping features between the entities[4]. The diagnosis of PNET is based on the histological and immunohistochemical staining of biopsy or surgical specimen; 90% of patients with PNET had a specific translocation gene t(11;22)(q24;q12) and the fusion gene EWS-FLI1 which could be found by Fluoroscence in situ hybridization(FISH) and reverse transcripstion polymerase chain reaction(RT-PCR) which can help in diagnosis[5]. As our patient was poor so molecular studies could not be undertaken.

Case Report

A 21 yr old male presented to a local hospital with complaints of intermittent flank pain and hematuria on and off. He had no history of any major illness or documented malignancy in the past. Ultrasonography identified a large lobulated left renal neoplastic mass, extending into proximal renal vein and collecting system. CT scan showed an exophytic heterogeneously enhancing solid soft tissue density mass lesion of size 8.6x8x6.4 cms arising from left kidney compressing left renal artery and distorting pelvi-calyceal system with provisional diagnosis of left side renal cell carcinoma. Positron enhanced tomography-CT revealed a large hyper metabolic soft tissue exophytic mass lesion arising from mid pole of left kidney infiltrating the lateral abdominal wall and renal pelvis with renal vein thrombosis suggestive of malignant etiology. No evident hyper metabolic distant metastases were noted elsewhere in the body.

Viewing the clinical and radiological profile, left radical nephrectomy was done and the surgical specimen was received in our department for histopathological examination. On gross examination, the specimen consisted of kidney covered partially with perinephric fat and ureter. On sectioning, a greyish brown mass of size 10x8x7 cm occupying all 3 poles of the kidney was noted with areas of necrosis and hemorrage. Renal sinus and capsule appeared invaded by the tumor grossly. Required sections were taken and processed. Histologically, tumor composed of small uniform, dark, round cells arranged in sheets and rosettoid pattern(Image 1). Cells had round to oval nuclei, dark clumped chromatin, inconspicuous nucleoli, and a small amount of vacuolated cytoplasm. Tumor cell infiltration was noted in renal capsule, peri-renal fat, renal sinus fat, gerota's fascia and renal vein. Hence on histomorphological basis it was reported as small blue round cell tumor suggestive of primary PNET left renal mass.

A wide panel of IHC markers were applied to rule out the other members of small round cell "blue tumor" family. Tumor cells showed strong diffuse membranous positivity for CD99 (Image 2) and moderate positivity for FLI1(Image 3). However tumor cells were negative for WT1, TLE1, Vimentin , LCA, TdT and Chromogranin. Thus the diagnosis was confirmed as renal ES/PNET and patient was further referred to department of medical oncology for initiation of treatment.

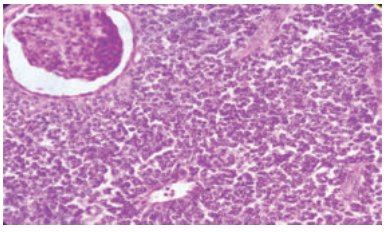

Figure 1. H&E Stain At 400x Magnification Showing Cells With Small Round Blue Nuclei With Finely Dispersed Chromatin And Scant Ill-defined Cytoplasm. Psedo-rossett Is Also Seen.

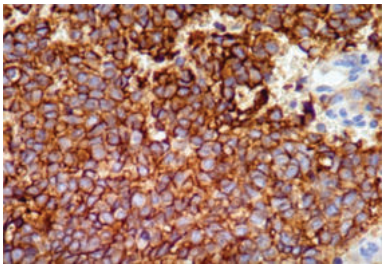

Figure 2. IHC Showing Diffuse Membranous Positivity Of Tumor Cells for CD99(400x)

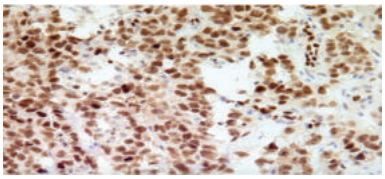

Figure 3. IHC Showing Nuclear Positivity Of Tumor Cells For FLI-1(400x)

DISCUSSION

Ewing's sarcoma (ES) and PNETs were formerly classified as two distinct pathological entities, albeit they both share common stem cell precursor and distinct chromosomal abnormality. Owing to their similar histologic and cytogenetic characteristics, these tumors are now categorized under a spectrum of neoplastic diseases now known as Ewing's sarcoma family tumors (ESFTs)[6].

Primary ES/PNET of the retroperitoneum, pelvis, paraspinal region and visceral organs like bladder, prostate, pancreas, ovaries etc have been described in the literature[7,8]. The first case of primary ES/PNET of the kidney was described by Seemayer and colleagues in 1975. Several theories explaining the origin of ES/PNET at peripheral sites have been proposed, such as presence of neural crest cell at these sites, tumor originating from neural ramification of celiac plexus innervating the kidney, derivation from the pleuripotent germ cells and/or from mesenchymal stem cells[8,9].

Renal ES/PNET usually presents with non specific symptoms like those of “classic renal tumor triads”; hematuria, pain and palpable abdominal masses. Due to highly malignant nature, ES/PNET progresses rapidly and invades the renal collecting system early leading to hematuria. It usually occurs in young adults, most often between 20 and 30 years with male:female ratio about 3:1[10]. Fewer than 200 cases are reported in the literature until now[2].

The first challenge for this kind of lesion is the diagnosis. The imaging feature of primary Extraosseous Ewing's sarcoma(EES) on contrast enhanced CT or Magnetic resonance imaging(MRI) are bulky heterogeneous masses with frequent local invasion or with compression of adjacent organs. Retroperitoneal masses are usually large at diagnosis with 50% of lesions bigger than 20cm[11].

The definitive diagnosis of renal ES/PNET is based on histopathology and Immunohistochemistry of resected specimens[10, 11]. Microscopically on H & E staining, renal PNET is characterized by monotonous proliferation of immature and small round cells. Formation of the Homer Wright rossets is typical feature of renal PNET[12]. The differential diagnosis on histology of ES/PNET of kidney includes the blastemal variant of Wilm's tumor, lymphoblastic lymphoma, Monophasic synovial sarcoma, solid variant of alveolar rhabdomyosarcoma, clear cell sarcoma of the kidney, neuroblastoma, desmoplastic small round blue cell tumor, small cell variant of osteosarcoma and small cell carcinoma[7,9]. Immunohistochemically, Renal PNET is frequently positive for CD99, a vital clue for the diagnosis. In addition, almost two-third reveal FLI1 expression. Other markers such as vimentin, cytokeratin, NSE and S100 have also been detected. Cytogenetic study of renal PNET is supportive to make the final diagnosis. Chromosomal translocation t(11;22)(q24;q12) usually can be identified in renal PNET resulting in formation of EWL/FLI1 fusion protein[12]. As our patient was poor so molecular studies could not be undertaken.

Renal ES/PNET treatment includes a multimodality approach including radical nephrectomy followed by combination chemotherapy with or without radiotherapy. Follow ups with laboratory and imaging tests are essential to assess recurrence and metastasis[13].

CONCLUSION

In general, Renal PNET is a rare malignancy that should be kept in the differential diagnosis of a renal space occupying lesion especially when it is presenting feature in adolescent and young adults. CT and MRI findings are non –specific but they are useful methods for local assessment of resectability and detection of metastases. However, the final diagnosis is based on histology and IHC which could be supported by molecular pathologic findings. The treatment is multimodal in nature but the prognosis remains poor. We expect that further awareness of this rare tumor can be established by presenting our case.

REFERENCES

1. Kumar V, Khurana N, Rathi AK et al. Primitive neuroectodermal tumor of prostate. Indian J Pathol Microbiol. 2008;51:386-88.
2. Liang L, Song H, Ma B, Zhang Z, Zhu K, Li Q et al. Renal Ewing's sarcoma/ primitive neuroectodermal tumor (PNET): a case series of 7 patients and literature review. Transl Androl Urol 2021;10(2): 548-54.
3. Pomara G, Cappello F, Cuttano MG, Rappa F, Morelli G, Mancini P et al. Primitive neuroectodermal tumor (PNET) of the kidney: A case report. BMC cancer 2004;4:3.
4. Randale AA, Parate SN, Bhatkule MA, Meshram SA, Tathe SP. Primary Ewing's sarcoma/primitive neuroectodermal tumor of kidney: A rare case report. Clin Cancer Investing J 2018;7:26-9.
5. Kang SH, Perle MA, Nonaka D et al. primary ewing's sarcoma/PNET of the kidney: Fine needle aspiration, histology, and dual color break apart FISH assay. Diagn Cytopathol 2007; 35:353-57.
6. Shangpliang DM, Rangad G, Das JK et al. Primary adult renal ewing's sarcoma; a rare entity. Cureus14(2): E22302.
7. Jagtap SV, Kale PP, Huddedar A, Hulwan AB, Jagtap SS. Primary primitive neuroectodermal tumor of kidney. Indian J Pathol Microbiol, 2018;01(2),282-84.
8. Gali SC, Kumarguru BN, Bhat B, Ramaswamy AS, Kumar MU. Renal primitive neuroectodermal tumor: Case report of a rare entity. J.clin Diagn. Res. 2017;11(2):ed17-19.
9. Kar S, Shah SN, Menon M, Geetha N. Primari Ewing's sarcoma/ Primitive neuroectodermal tumor of kidney; report of four cases from a tertiary care center. CPCCR 2021:3;100043.
10. Zhang S, Li Y, Wang R, Song B. Ewing's sarcoma/primitive neuroectodermal tumor of the kidney: A case report and literature of review. Transl Androl Urol 2019;8(5):562-66.
11. Cochetti G, Paladini A, Vermandois JAR, Fatigoni S, Zanelli M, Ettore M et al. Metastatic renal Ewing's sarcoma in adult woman: Case report and review of literature. Open med (wars). Mar 9;16(1):397-409.
12. Yang C, Xu H, Zhou J, Hao Z, Wang J, Lin C et al. Renal primitive neuroectodermal tumor: A case report. Medicine:2015, Vol 94(49);e2304.
13. Alghamdi MHA, Alawad SA, Alharbi MG, Alabdulsalam AK, Almodhen F and Alasker A. A rare case of ewing's sarcoma of the kidney. Urol case reports 2020: vol29;101094.