



RESEARCH ARTICLE

PRIMITIVE NEUROECTODERMAL TUMOUR OF KIDNEY: A RARE CASE REPORT

*Kaushal Rai and Dr. Manjusha Madhusudan Litake

B.J.G.M.C. Pune, India

ARTICLE INFO

Article History:

Received 14th February, 2017
Received in revised form
19th March, 2017
Accepted 17th April, 2017
Published online 23rd May, 2017

ABSTRACT

Primitive Neuro ectodermal tumors (PNET) are predominantly a childhood tumor of ewing's sarcoma family with very aggressive nature. Most common site of involvement includes CNS and skeleton system. Renal PNET is a rare, extra-cranial entity. Characteristic histological features of these tumors are small, uniform round, blue cells. Differential diagnosis of PNET of kidney includes lymphoma, monophasic Wilm's tumor, carcinoid, clear cell sarcoma. Here presenting a rare case of primitive neuro ectodermal tumor of kidney in a 12 year old female.

Key words:

Primitive Neuroectodermal
Tumor (PNET);
Ewing's sarcoma;
Wilm's tumor.

Copyright©2017, Kaushal Rai and Dr. Manjusha Madhusudan Litake. This is an open access article distributed under the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.

Citation: Kaushal Rai and Dr. Manjusha Madhusudan Litake, 2017. "Primitive neuroectodermal tumour of kidney: a rare case report", *International Journal of Current Research*, 9, (05), 50608-50610.

INTRODUCTION

A 12 year old female child was admitted with complaints of pain in left hypochondriac and lumbar region in abdomen since 3 months, with increased intensity of pain for 3 days, with no urinary complaints. Physical examination revealed a palpable lump in left hypochondriac region of firm consistency with regular margin, crossing midline with mild tenderness. Ultrasonography of abdomen was suggestive of 12.5×10.1×8.9 cm, well defined heterogeneous mass arising from upper pole of left kidney with splaying of calyceal system with mild vascularity. CT scan revealed well-defined 12.1×10.3×12 cm, exophytic, heterogeneously enhancing, soft tissue mass lesion arising from upper and mid pole of left kidney, extending from T9 to L3 vertebra, displacing pancreas, bowel loops. This lesion was encasing the left renal artery and also causing compression of left renal vein with its non-opacification. Left adrenal gland was not separately visualized from the above lesion and remaining renal parenchyma was compressed with obstruction of pelvic calyceal system, with no evidence of significant lymphadenopathy. Aorta and inferior vena cava were normal. Left sided Radical nephrectomy was performed with left thoraco-abdominal incision under general anesthesia.

Intra- operative finding suggestive of solid and cystic tumor arising from left kidney, of size approx. 15×10×10cm and merged with Gerota's fascia and without distinct separation of renal parenchyma and adrenals from tumor mass. Tumor mass was friable at some places. Renal vessels separately identified entering in the tumor mass. Multiple lymph node enlargements along the abdominal aorta, were dissected and sent for histopathology. Macroscopically specimen measured 12×10×10 cm weighted 350grams with involvement of whole left kidney, at some places in the periphery of the tumor normal kidney is identified with areas of necrosis and hemorrhage. At places small cystic areas seen with no lymph node identified. On microscopic view shows tumor mass composed of sheets of round cell with hyperchromatic nuclei with scanty eosinophilic cytoplasm, at places nuclei are eccentrically placed giving plasmocytic appearance. Sheets are separated by fibrous septa with no lymphoid tissue seen suggestive of malignant round cell tumor with differential of Neuroblastoma and Non-Hodgkin's lymphoma.

Histopathology

In view of inconclusive histopathology report, Immunohistochemical staining was done (BiogenexEZ Retriever in Tris-EDTA buffer PH 9.0). Tumor cells were focally positive for CD99 (Clone HO36.1.1) and Fli-1 (Clone MRQ1) and negative for LCA (Clone LCA88), Synaptophysin (Clone

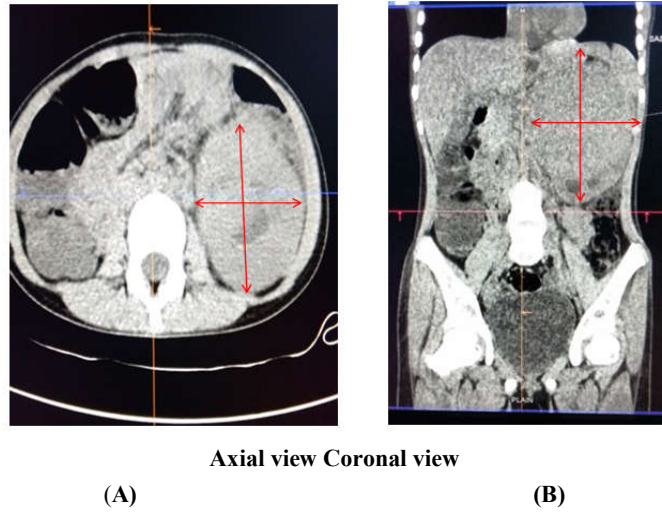


Fig 1.0. Computed tomogram of abdomen showing a well-defined tumor involving whole of the left kidney, filling almost the entire left side of abdomen



Fig. 2.0 Gross specimen of the tumor

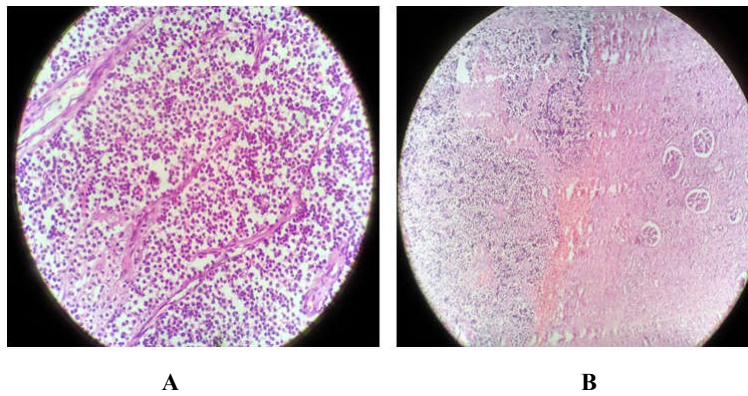
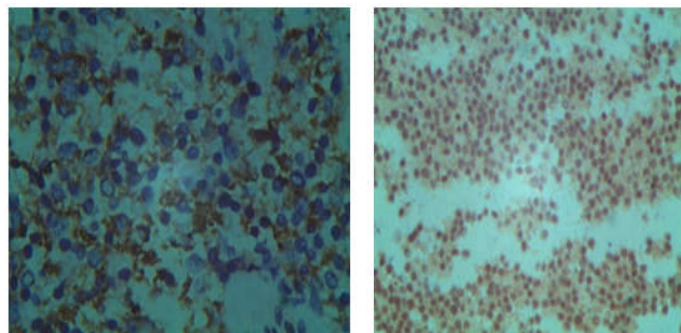


Fig:2.1 HE stain ,40x: (A) tumor cells with fibrous septa;(B) tumor with normal kidney



A: CD 9940XB: FLi-1 20X

Snp88), Chromogranin, Desmin (Clone 33) supported the diagnosis of Renal Primitive Neuroectodermaltumor. Oncology follow-up advised 2 course of chemotherapy, each consisting of 5 days therapy of Ifosfamide and etoposide. At the same time after one month of post-operative period, repeat computed tomogram of abdomen was suggestive of multiple masses in retroperitoneum, in left paravertebral region involving posterior to body and tail of pancreas as well as recurrence and metastatic deposit in left side of rectus. Post chemotherapy repeat computed tomography of abdomen and pelvis was done at 3 months interval suggestive of heterogeneous mass lesion of sized 5.8×2.8×2.6 cm present in left renal fossa below spleen with no mass lesion in paravertebral and rectus muscle. Due to invasion to paravertebral region resulting excruciating pain, palliative pain block was given. However patient succumbed post-operatively within six months due to rapid and aggressive recurrence at multiple sites.

DISCUSSION

In 1918, Arthur Purdy Stout described a tumor composed of small round cells with rosettes, in the ulnar nerve, which came to be known as primitive neuroectodermal tumor (PNET) (Azar, 1998). These represent less than 1% of all sarcomas, predominantly affects bones and deep soft tissue, rarely affects visceral organs. The incidence of PNETs in the body as follows the thorax (44%), abdomen and pelvis (44%), extremities (20%) and head and neck region (6%). Primarily visceral involvement with decreasing frequency are pancreas, vagina, stomach, small bowel, ovaries, esophagus and kidney (Mallén Mateo *et al.*, 2005). In year 1994, Mor described the first case of renal PNET. Other differential diagnosis of PNET of kidney includes lymphoma, monophasic Wilm's tumor, carcinoid, clear cell sarcoma (Habermann, 2003). Patients usually present with non-specific symptoms like discomfort, pain and a palpable mass on examination. For definitive diagnosis biopsy is essential. On histopathology, gross appearance in general, is multilobulated, soft, and friable. Cut section shows gray-tan or gray-yellow appearance with large areas of necrosis and hemorrhage and calcification is rare (Ellinger *et al.*, 2006). On microscopy typical PNET shows lobular growth pattern with little stroma composed of poorly differentiated, dark stained round cells. Many PNETs on immunohistochemistry positive for CD99, neuron specific enolase, S-100, Leu-7 and synaptophysin and negative for desmin and myogenin. In some cases they are positive for cytokeratin (Chung *et al.*, 1998; Doi *et al.*, 2009). For extraskeletal PNET, treatment with surgery alone is considered insufficient, and multimodal treatment with chemotherapy and radiotherapy is performed. In spite of this multimodal treatment, there is rapid dissemination of the disease seen in most of the patients (Ellinger *et al.*, 2006). The outcome of the treatment at the time of diagnosis is adversely affected by various prognostic factors are large tumor size, extensive necrosis, poor response to initial chemotherapy as seen in presented case (Ellinger *et al.*, 2006; Kuroda *et al.*, 2000). Most commonly used chemotherapeutics are adriamycin, etoposide, dactinomycin, vincristine, cyclophosphamide, and ifosfamide (Grier *et al.*, 2003).

In presented case diagnosis of primary neuro ectodermal tumour of kidney was confirmed by hystopathology and immuno-histochemistry. Immuno-histochemistry particularly helpful, showing positivity for CD99 and negative for desmin and synaptophysin.

Conclusion

Renal PNET is rare and aggressive tumor of which diagnosis and treatment is challenging. Immuno-histochemistry plays great role in diagnosis of renal PNET. Recurrence is common after surgical removal and tumour is chemosensitive, so post-op adjuvant chemotherapy should be recommended. Even though rPNET is rare it should be considered in differential diagnosis of renal masses.

REFERENCES

- Azar, H. A. 1998. Arthur Purdy Stout (1885-1967), a pioneer of surgical pathology: a survey of his Notes on the Education of an "Oncological" Surgical Pathologist. *Annals of Diagnostic Pathology*, 2(4), 271-279.
- Chung, D. H., Lee, J. I., Kook, M. C., Kim, J. R., Kim, S. H., Choi, E. Y., ... Song, H. G. 1998. ILK (beta1-integrin-linked protein kinase): a novel immunohistochemical marker for Ewing's sarcoma and primitive neuroectodermal tumour. *Virchows Archiv: An International Journal of Pathology*, 433(2), 113-7.
- Doi, H., Ichikawa, S., Hiraoka, A., Ichiryu, M., Nakahara, H., Ochi, H., ... Hasegawa, T. 2009. Primitive neuroectodermal tumor of the pancreas. *Intern Med*, 48(5), 329-333.
- Ellinger, J., Bastian, P. J., Hauser, S., Biermann, K., & Müller, S. C. 2006. Primitive neuroectodermal tumor: rare, highly aggressive differential diagnosis in urologic malignancies. *Urology*, 68(2), 257-62.
- Ellinger, J., Bastian, P., Hauser, S., Biermann, K., Müller, S. 2006. Primitive neuroectodermal tumor of the kidney. *Der Urologe. Ausg. A*, 45(6), 735-738.
- Grier, H. E., Krailo, M. D., Tarbell, N. J., Link, M. P., Fryer, C. J., Pritchard, D. J., Miser, J. S. 2003. Addition of ifosfamide and etoposide to standard chemotherapy for Ewing's sarcoma and primitive neuroectodermal tumor of bone. *The New England Journal of Medicine*, 348(8), 694-701. <http://doi.org/10.1056/NEJMoa020890>
- Habermann, H., Beneschb, M., Schipsa, L., Pummera, K., Ratschekc, M., Uggowitzerd, M.M., Urbanb, C., Hubmera, G. 2003. Findings and clinical course of a localized primitive peripheral neuroectodermal tumor of the kidney. *Urol Int*. 71:319-321. doi: 10.1159/000072686.
- Kuroda, M., Urano, M., Abe, M., Mizoguchi, Y., Horibe, Y., Murakami, M., Kasahara, M. 2000. Primary primitive neuroectodermal tumor of the kidney. *Pathol.Int.*, 50(1320-5463 (Print)), 967-972.
- Mallén Mateo, E., Sancho Serrano, C., Pascual Regueiro, D., García de Jalón Martínez, A., Gil Sanz, M. J., Gonzalvo Ibarra, A., & Rioja Sanz, L. A. 2005. [Primitive neuroectodermal tumor. Ewing's sarcoma]. *Actas Urológicas Españolas*, 29(3), 311-3.
