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CASE REPORT

Aggressive Primary Neuro Ectodermal Tumour in Kidney: A Rare Entity

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ABSTRACT

Primitive Neuro-Ectodermal tumor (PNET) of kidney is a rare tumor with only a few published reports. We report here a case of PNET of kidney in a female aged 20 years who reported of vague pain and lump in loin with a history of rapid increase in size of the lump. On CT imaging, on the left side a large heterogeneous, enhancing mass 15.1x15.4cmx17cm was seen arising from the upper pole of the left kidney.Radical nephrectomy was done along with removal of para-aortic nodes detected intra-operatively. Histopathology revealed sheets of small round cells intervened by hemorrhagic cystic areas with surrounding thin rim of normal kidney tissue. Immunohistochemistry showed diffuse membrane positivity of tumor cells for CD99. Post-operative USG carried out 18 weeks after surgery revealed an ill-defined retroperitoneal mass with ascites, pulmonary metastasis and pleural effusion with presentation of sub-acute intestinal obstruction. The patient succumbed to pulmonary complications later. PNET of the kidney is a very rare and aggressive tumour with poor prognosis. The disease-free survival rate at 7.5 years is around 45–55% in well-confined cases.

***Keywords:** Primitive neuro-ectodermal tumor, round cell tumour, ewings sarcoma, rare renal tumor*

INTRODUCTION

Primitive Neuroectodermal Tumor (PNET) of kidney is a rare tumor with only few published reports. This tumor is the soft-tissue equivalent of the Ewing sarcoma and malignant small cell tumor of the thoraco-pulmonary region.[1] Primitive neuroectodermal tumor is a type of sarcoma that occurs in the first two decades of life.[2] As the tumor is highly aggressive, it is often diagnosed in advanced stage.[3] We report here a case of Primitive Neuro-Ectodermal Tumor of kidney in a female aged 20 years

CASE HISTORY

An unmarried young female presented with a month long history of vague abdominal pain. She noticed fullness of the flank and a gradually progressing distinct swelling on her left upper abdomen with dull pain initially, radiating to back. During the course of the last one-month there was sharp increase in intensity of pain and she sought medical help when over the counter drugs failed to offer

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relief. She had no fever, chills, nausea, vomiting, diarrhoea, or change in bowel habits. She had no history of hematuria, dysuria, pain or burning on urination, bloody stools or any other significant abdominal ailment in the past. Her menstrual history too was insignificant.

On physical examination her blood pressure was 110/70 mm Hg, pulse was 96 beats per minute, respirations were 18/min and temperature was 37.3°C. Her general examination was unremarkable. Her heart sounds were regular and without murmurs, and a pulmonary examination showed equal breath sounds bilaterally. Abdominal examination revealed an irregular, fixed, tender mass in the left upper quadrant, filling the entire left upper quadrant and palpable upto the pelvic rim. The mass was approximately 15 cm long, extending from the left subcostal margin to the anterior superior iliac crest on the left side and crossed the midline. The inferior border of the mass was ill defined and difficult to follow.

Her laboratory values were as follows: urinalysis was negative for any red blood cells, white blood cells, bacteria, glucose, or protein and the culture report was sterile. Her white blood cell count was 9,400/µL, with hemoglobin of 7.7 g/dL, and a hematocrit of 37.7%. Renal function test: creatinine 0.8 mg/dL, blood urea nitrogen 16 mg/d, sodium 139 mEq/L, potassium 4.1 mEq/l and glucose 90 mg/dL. CECT of the abdomen showed 15.1cm x 15.4cm x 17cm well defined isodense mass arising from the upper pole of the kidney with heterogenous enhancement and few areas of hypo-density suggestive of necrosis.

She was posted for radical nephrectomy and during surgery, an 18x16x16cm tumor was found invading beyond the Gerota's fascia. Histopathological examination showed extensive necrosis and hemorrhage within the tumour with small round cells arranged in sheets suggestive of round cell tumour. On immunohistochemistry it stained positively for CD 99. We could not do fluorescent in situ hybridization to demonstrate EWS-FLI-1 gene fusion. Tumour cells were found in 7 of the 11 nodes removed. The patient recovered well after surgery and was discharged on the 10th post-operative day with the advice to attend after 2 weeks for chemo-radiation. However, she failed to turn up for follow up and was readmitted 18 weeks later with a mass of 4cmx6cmx6cm in the renal bed, which was encasing the aorta. This mass also impinged on her left colon. She presented with features of sub acute intestinal obstruction. Her chest radiographs showed bilateral multiple nodular opacities suggestive of metastasis. Her condition worsened with severe pulmonary and gastro-intestestinal symptoms. She succumbed to pulmonary complications, two weeks after her admission.

DISCUSSION

PNET belongs to the Ewings family of sarcomas and bears pathological similarity to its bony lesion.[1, 4, 5] They were first described by Stout in association with peripheral nerves.[6] PNET shows strong positivity for MIC-2 gene product like CD-99 by which it can be distinguished from other small round cell tumors. The distinguishing genetic factor in primitive neuroectodermal tumor is the association with a translocation between chromosomes 11 and 22, the t(11;22)(q24;q12).[7, 8, 9] If needed, the diagnosis can be confirmed by demonstration of the t(11:22) or the EWS-FLI-1 gene fusion with the help of fluorescent in situ hybridization (FISH) technique or RT-PCR which was not done in our case.[2, 10] In our case diagnosis of primitive neuroectodermal tumor was based on the histopathological findings of the surgical specimen and immunohistochemical staining. The National Wilms' Tumor Study Group in their study have concluded that these malignancies represent a diverse group of high-grade tumor which is not always easy to place in a single category, even though they are evaluated immunohistochemically and by molecular genetic tools.

Children and adolescents are most frequently affected. The most common locations are the head, neck, trunk, and extremities.[6] The clinical features of PNET kidney may be nonspecific like vague pain and lump in the left loin as seen in our case and there may be irregular fever, weight loss, and occasional hematuria. The differential diagnosis of renal tumors in this age group includes renal cell tumor, Wilms tumor, and lymphoma.[7, 8]

The treatment of primitive neuroectodermal tumor is surgical resection and an adjuvant chemotherapy regimen consisting of combinations of doxorubicin, cyclophosphamide, vincristine, and dactinomycin. In some cases chemotherapy is started before surgery.[8] Radiation therapy may prove beneficial in some cases. These tumors have a poor prognosis, with a disease-free survival of 45% at 7.5 years.[7] In view of its poor prognosis and aggressive nature renal PNET should be differentiated from other small blue round cell tumors like neuroblastoma,

rhabdoid tumor of kidney, nephroblastoma, small cell carcinoma, synovial sarcoma (monophasic, poorly differentiated) and non-Hodgkin lymphoma (NHL).

A review of the literature revealed that there are very few cases of PNET involving the kidneys reported. The diagnosis of such tumours is based on the clinical setting of aggressive nature of the tumour growth in the adolescent age group and they need early surgical extirpation. A prompt diagnosis suggested by immunohistochemistry and early chemotherapy regimen may be helpful in such a situation to improve the prognosis.

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