University Journal of Surgery and Surgical Specialties

ISSN 2455-2860 Volume 2 Issue 3 2016

Primitive Neuroectodermal Tumor - Kidney A Case Report

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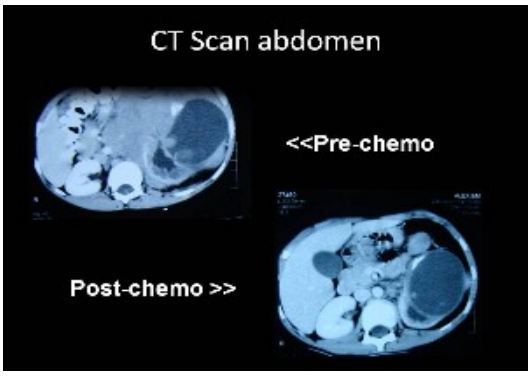
Abstract :

Peripheral primitive neuroectodermal tumors are rare tumors representing 1percent of all sarcomas and renal localization is very rare (intra abdominal-14 percent). We are reporting a case of PNET of left kidney. The relevant literature has been reviewed and a brief discussion on PNET kidney has been added.

Keyword :primitive neuroectodermal , tumor , kidney , PNET

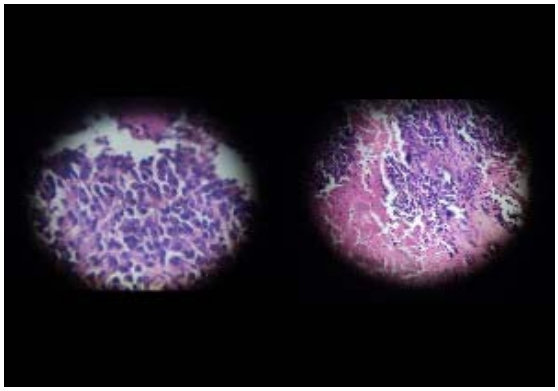
Introduction:



Primitive neuroectodermal tumor of kidney is classified as a malignant tumor and has an aggressive behavior **Case report** 9 years old boy with a huge mass left flank mass crossing the mid line . 2 months ago child underwent native treatment for hepatitis and one episode of hemoptysis. On examination there were no features to suggest syndromic renal tumors. BP was normal. Per abdomen - A bosselated mass of varying consistency was found occupying the entire left flank. A large varicocele was present on the left side .Hernial orifices were free. Both testis descended. On investigation - renal function tests were normal.Plain X-ray abdomen shows a soft tissue shadow pushing the bowel loops to the right side . USG shows a left kidney mass 10 × 15×13 cm with areas of calcification and necrosis. Tumor markers -AFP , B-HCG , VMA were negative. Bone marrow was negative. X-ray skeletal survey was normal.

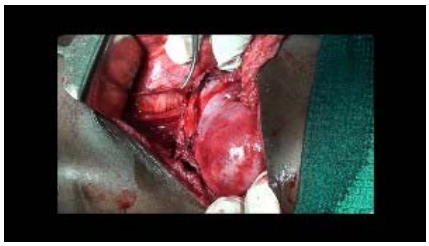
CECT showed evidence of tumour originating from left kidney with great vessels pushed to right.

A tumour of left renal origin was suspected (wilms /non wilms). An open biopsy by extraperitoneal approach was done .HPE showed small round cells tumor. Immuno histochemistry panel was positive for CD99 , Vimentin and negative for WT1 ,Synaptophysin and chromogranin. So with the provisional diagnosis of PNET- VAC/IE regime was started. On follow up by USG & CECT showed regression in mass size with great vessels back to normal position. Left nepheroureterctomy was done. HPE –L Kidney shows chemotherapy related changes,round cells with pseudo rosettes, capsule negative for tumor . post operatively 4th cycle of chemotherapy was given followed by radiotherapy. Repeat USG shows no mass ,R kidney is normal.IHC panel was positive for CD99,WT1,NSE 2+ve.



HPE – Small round cells tumor

Excised Tumor



Per-operative picture

Discussion:

Peripheral primitive neurectodermal tumor was first recognized by Arthur Purdy Stout[1] in 1918. It belongs to the family of small round cell tumors.Renal site is very rare and 50 cases have been reported in literature .Exact no of cases not known as it has often not been differentiated from Ewing's sarcoma.Renal PNET is more aggressive than in other locations .It arises in childhood or adolescsnce and has an aggressive course towards metastatic disease and death and recurs locally .It metastasis early to regional lymph nodes,lung , liver, bone , bone marrow leading to poor prognosis. 5yr disease free survival rate for patients for patients with extraskeletal [2] PNET is 45-55% Differential diagnosis includes- extra osseous ewings sarcoma, rhabdomyosarcoma, wilms tumor ,carcinoid,neuroblastoma,clear cellsarcoma of the kidney,lymphoma,small cell variant of osteosarcoma desmoplastic small round cell tumor . Homer – wright type rosettes are typical histological features[3] of primitive neuroectodermal tumors.Immunohistochemical analysis is necessary for diagnosis.Treatment for renal PNET is surgery , chemotherapy, radiotherapy.Even with aggressive treatment prognosis remains poor.Advanced

disease at presentation has a relapse free survival of only 2yrs.

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