ORIGINAL RESEARCH PAPER

Volume-8 | Issue-8 | August - 2019 | PRINT ISSN No. 2277 - 8179

INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

PRIMARY EWING SARCOMA OF THE KIDNEY, A RARE PRESENTATION: A CASE REPORT

General Surgery

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ABSTRACT

Primitive Neuroectodermal Tumour (PNET) is a small round cell malignancy of primitive neuroectodermal tissue or migratory, pluripotent neural crest cells that arises from the soft tissue (extra skeletal variety) or bone, affecting older children and adults. Recently, Ewing sarcoma and PNET have been unified into a single category: the ESFT (Ewing Sarcoma Family Tumours) based on shared clinical, morphologic, biochemical and molecular features [1]. In this article, we present here a case of a 35-year old male with metastatic Ewing sarcoma of the left kidney which is a very rare entity.

KEYWORDS

Primitive Neuroectodermal Tumour Kidney, Ewing Sarcoma Kidney, Nephrectomy, Debulking

PRESENTATION OF CASE

A 35-year-old male presented with chief complaints of dull-aching pain in the left loin accompanied by the passage of blood in urine and mild respiratory discomfort for the past 1 week. On clinical examination, a palpable ballotable left loin mass was found, suggestive of a renal mass. All blood parameters were normal. Sonography of the abdomen revealed a large heterogenous lobulated mass not separate from the left kidney. Doppler revealed hyper vascular lesion. Chest roentgenogram was normal. No tumour thrombus in the renal vein and inferior vena cava.

The patient underwent a contrast-enhanced computed tomography of the abdomen and pelvis for the suspected renal lesion, which revealed a large lobulated mass lesion in the left kidney, replacing almost the entire left kidney, with vertebral osteolytic lesions suggestive of bony metastases. Patient had para-aortic and pre-aortic lymph node enlargement. The contralateral kidney showed prompt excretion of contrast and good function. In view of persistent haematuria, and suspected malignant lesion of the left kidney (based on imaging), the patient underwent a left-sided radical nephrectomy.

Intra-operatively, the kidney was entirely replaced by the tumour tissue which was hyper- vascular. Left sided radical nephrectomy was carried out with control over the renal pedicle. All the lymph nodes (pre- and para-aortic, till the aortic bifurcation) were removed along with the specimen. The patient had uneventful postoperative recovery, was discharged on post-operative day 9, and was asked to follow-up.

Histopathology of the tumour revealed small round blue cell tumour suggestive of Ewing sarcoma of the left kidney. In the subsequent follow-up visits, the patient deteriorated rapidly within 2 months and developed altered sensorium suggestive of cerebral metastases. Patient developed generalized weakness, debility, multiple palpable metastatic lesions in the skull and at the end of 2 months, patient succumbed to death despite of all attempts to revive.

Considering the age of the patient, Wilms tumour and renal cell carcinoma were unlikely differential diagnoses. Tumour of unusual cell origin was suspected and expectedly turned out to be PNET(Ewing sarcoma).

PATHOLOGICAL FEATURES

Histopathological examination revealed a unifocal tumour involving the renal sinus but sparing the perinephric fat, Gerota fascia, renal artery and vein, pelvicalyceal system, ureter. There was presence of a high-grade malignant round cell tumour with intervening thin-walled vessels. Margins were free of tumour and no lymphovascular invasion or rhabdoid features could be appreciated. The final histopathological reporting was suggestive of Ewing sarcoma/PNET of the kidney, showing the following features on the immunohistochemical staining pattern, which were indeed consistent with the diagnosis of Ewing sarcoma/PNET of the kidney:

1. membranous diffuse positivity for CD-99 i.e. MIC2
2. weak and focal staining for FLI1
3. negative for synaptophysin, WT1, CD-56, chromogranin

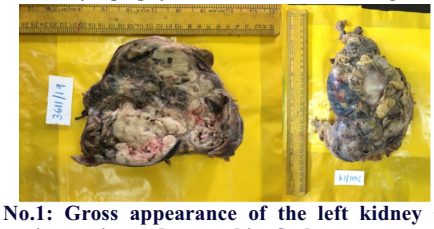


Figure No.1: Gross appearance of the left kidney specimen demonstrating variegated grey-white fleshy tumour mass, which has almost replaced the entire kidney.

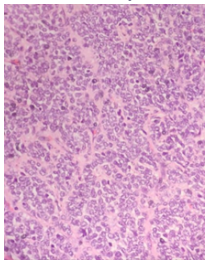


Figure No.2: 100x magnification with Haematoxylin and Eosin staining, demonstrating the tumour composed of malignant small round cells all over the field.

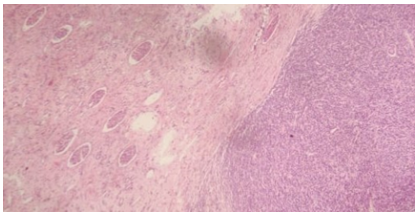


Figure No.3: 100x magnification with Haematoxylin and Eosin staining of the tumour tissue demonstrating the histologically normal renal parenchyma on the left, which is invaded by the small round blue cell tumour on the right.

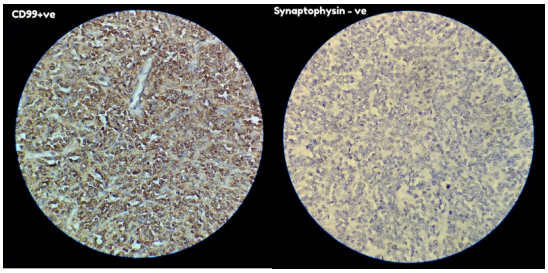


Figure no.4: 400x magnification with immunohistochemical staining demonstrating CD99 positivity, which is almost pathognomonic of Ewing sarcoma. The tumour also demonstrated Synaptophysin negativity.

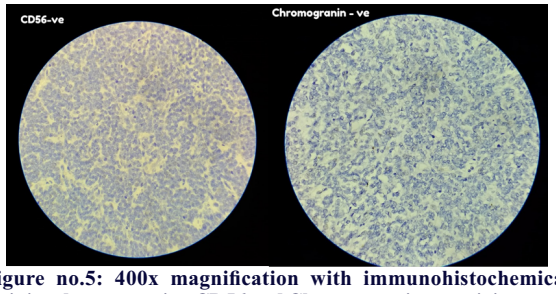


Figure no.5: 400x magnification with immunohistochemical staining demonstrating CD56 and Chromogranin negativity.

DISCUSSION:

Ewing sarcomas are extremely rare tumours of the kidney which are seen in young age group where RCC is uncommon. Angioma, angiomyolipoma, tumours of the adrenal gland are common after the age of 25 years. When the tumours are involving the whole kidney, a nephrectomy becomes mandatory. It is not routine practice to do pre-operative tissue diagnosis in renal tumours. The decisions are based on radiological imaging features, presence of hyper or hypo-vascularity, functional status of the contralateral kidney.

Computed tomography revealed the following findings:

- A mass lesion replacing almost entire the left kidney; extending from the level of D11 to L3 vertebral bodies showing heterogeneous post contrast enhancement and areas of necrosis. These findings were suggestive of malignant neoplastic aetiology like sarcomatoid renal cell carcinoma more than transitional cell carcinoma of renal pelvis. Lymph nodal and bony metastases were present.

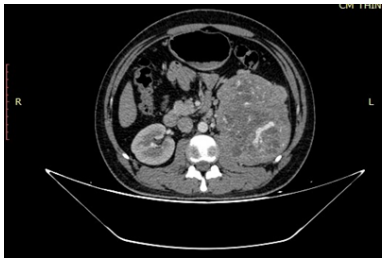
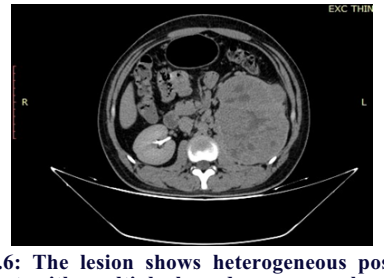


Figure No.6: The lesion shows heterogeneous post contrast enhancement with multiple hypodense non-enhancing areas within suggestive of necrosis. There is loss of appreciation of normal anatomy of left kidney with extension of the lesion in the left renal pelvis and along the upper ureter.



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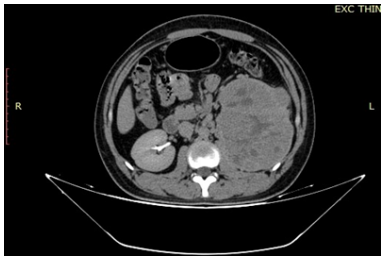

Figure No.7: No excretion of contrast is noted even on delayed (7 hour) scan from left kidney.

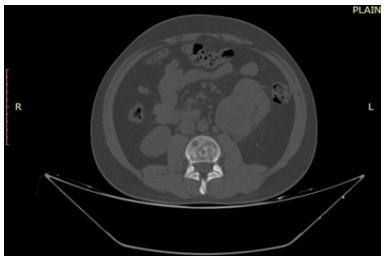

Figure No.8: Osteolytic metastatic lesion noted in the body of the dorsolumbar vertebral bodies.

FINAL DIAGNOSIS

Primary Ewing sarcoma (EWS) of the kidney is a rare tumour in adults. It was first described in 1975 by Seemayer and colleagues, and has since been sporadically documented in the literature [2]. PNET/ESFT (Ewing Sarcoma Family of Tumours) of the kidney are relatively rare but aggressive neoplasms which have a tendency to metastasize early, as evidenced in this case report. Clinical presentation may range from an asymptomatic loin mass, to a palpable mass with symptoms of haematuria, flank pain and weight loss. Based on the findings of histopathology, immunohistochemistry and computed tomography images we have arrived at the diagnosis of Primitive Neuroectodermal Tumour of the kidney.

Ewing Sarcoma was first described by Stout in 1918. Ewing sarcoma was recognized by Stout to contain small round blue cells arranged in rosettes [3]. Renal EWS is an extremely rare entity, which has been documented throughout the literature as isolated case reports. It has been reported that most patients presenting with primary EWS of the kidney present at a median age of 28 years. It carries a slight preponderance in men, and a worse prognosis if metastases are present at the time of diagnosis [4]. Approximately, 85–90% of EWS can be defined by a DNA translocation t (11;22) (q24; q12) [5,6].

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