INTRODUCTION

Ewing's sarcoma, also called Primitive Neuro Ectodermal Tumour (PNET), belongs to a family of small round-cell tumours known as The Ewing family of tumours. A common genetic locus is responsible for a large percentage of Ewing's sarcoma (ES) and primitive neuroectodermal tumours (PNET). They are grouped together as the Ewing's sarcoma family of tumours. It is usually seen in children and adolescents. Ewing's sarcoma is a rare disease usually classified as bone tumour, although it can have characteristics of both mesodermal and ectodermal origin, making it difficult to classify. Renal Ewing's sarcoma is even rarer in children.

Here, we report the case of a 6 years old girl who presented with abdominal mass and posed much diagnostic challenge. The patient underwent complete surgical excision of tumour, and is being treated with aggressive adjuvant chemotherapy.

CASE REPORT

A 6 years old girl child presented to the Haematology/Oncology Department of Children Hospital with 2 months history of abdominal distension. On examination, she was pale with stable vital signs and growth parameters on the 25th centile for age. Her abdominal examination revealed distended abdomen with 10 x 8 cm non-tender mass palpable in the left lumbar region reaching up till midline. Rest of the systemic examination was unremarkable. Ultrasound abdomen showed a 12.6 cm solid mass arising from the left kidney extending close to midline with some enlarged paraortic lymph nodes. Complete blood counts, serum electrolytes, liver function tests and chest X-ray were normal. Clinical suspicion of left sided Wilms' tumour was made and further workup done for better delineation of the tumour and distant metastasis. CT scan abdomen and chest showed a huge heterogeneous dense mass arising from left kidney, paraortic lymphadenopathy and multiple lung parenchymal nodules favouring metastatic Wilms' tumour.

Ultrasound guided trucut biopsy was done and suspecting the diagnosis of Wilms' tumour, 6 weeks pre-operative chemotherapy was started according to UKCCSG protocol. Histopathological examination showed a neoplasm composed of round blue cells having hyperchromatic nuclei, however, immunohistochemistry was advised. Immunohistochemical stains performed were diffuse strongly positive for MIC-2, However, WT1, NF1, Desmin, LCA, Cytokeratin AE1/AE3, TdT were all negative. These morphological and immunohistochemical features favoured Ewing's sarcoma/primitive neuroectodermal tumour (PNET).

The parents were counselled and further workup was planned for renal Ewing's sarcoma. CT scan chest and abdomen repeated showed 6.5 by 5.0 cm soft tissue mass arising from upper and mid pole of left kidney. However, there was no evidence of parenchymal abnormality in either lung. For evaluation of distant metastasis, bone scan and bilateral bone marrow aspiration biopsy were both normal. As there was slight reduction in size of the mass, surgical opinion was taken for resectability of the tumour. Left nephrectomy was...
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done on 15 April 2013. Postoperatively, patient had a smooth recovery. Chemotherapy has been started for Ewing’s sarcoma according to Euro Ewing Protocol. Till now child has completed three cycles of chemotherapy, is tolerating chemotherapy well and is on our regular follow-up.

**DISCUSSION**

Primary renal Ewing’s sarcoma/primitive neuroectodermal tumour (PNET) is an extremely rare neoplasm in children. Its usual age of presentation is in young adults and only a few paediatric cases have been reported.\(^1\)\(^2\)\(^3\) Renal PNET, as a separate entity, was reported first in 1975.\(^5\) Till now, only a small number of primary renal Ewing’s/PNET cases have been reported. Paediatric cases (younger than 15 years old) of ES/PNET of the kidney are extremely rare, and worldwide only 10 cases have been reported previously.\(^7\)\(^-\)\(^9\)

Ewing’s sarcoma/primitive neuroectodermal tumour (ES/PNET) is an extraordinarily rare primary tumour in the kidney and can be mistaken for a variety of other round cell tumours, including blastema-predominant Wilms’ tumour (WT). Approximately 90% of ES/PNET have a specific t(11;22), which results in a chimeric EWS-FLI1 protein. ES/PNET of the kidney needs to be differentiated from other small round cell tumours of the kidney because each type of tumour is treated differently. Ewing’s sarcoma/PNET has diagnostic genetical findings. The most common translocation is t(11;22) (q24;q12) with EWSR1-FLI1 genfusion (> 90%).\(^10\)\(^-\)\(^11\) Clear distinction is important between two entities as it has prognostic and therapeutic implications. To diagnose Ewing’s sarcoma, the combination of morphological findings, immunohistochemical analyses, and genetic changes together forms the base of the diagnosis.

The common presentation of extraosseous Ewing’s sarcoma is with a mass. In the index patient presenting with a unilateral solid renal mass Wilms’ tumour is usually the first differential followed by clear cell sarcoma of kidney, neuroblastoma and rhabdomyosarcoma. Clear cell sarcoma is distinguished by frequent skeletal metastases and neuroblastoma by its suprarenal origin and vessel encasement. Further delineation from other round blue cell tumours is made by distinct pathological changes and immuno-histochemistry. Histologically, Ewing’s sarcoma tumours comprise small round cells, that express CD99, and characterized by a specific chromosomal translocation involving the EWSR1 gene on chromosome 22, with an erythroblastosis virus-transforming gene (FLI1 in 85% of cases), resulting in a fusion oncogene.\(^1\)

Regarding prognosis, the 5-year disease-free survival rate of ES/PNET is 65 - 85% but the prognosis of ES/PNET of the kidney appears worse.\(^12\) Mukkunda et al.\(^13\) found in their analysis of 7 patients with renal Ewing’s sarcoma and with median follow-up of 36 months (range from 5 to 149), a median disease-free survival in patients with non-metastatic disease of 30.35 months (range from 5.1 to 149) with a 5-year overall survival rate of 42%.

Treatment of Ewing’s sarcoma is multimodal and requires multidisciplinary approach with involvement of paediatric oncologist, surgeon and radiologist. The role of radiotherapy is not well established. Chemotherapy lasts 6-9 months and consists of chemotherapeutic regimens containing vincristine, doxorubicin, cyclophosphamide/ ifosfamide and etoposide. It is important to differentiate Ewing’s sarcoma / primitive neuroectodermal tumour of the kidney from other common renal neoplasms because treatment options for the different types of renal tumours are vastly different and the need for a correct diagnosis is, therefore, crucial for appropriate patient management and counselling.

**REFERENCES**


