CASE REPORT

Primary Ewing's sarcoma of the kidney presenting with left-sided varicocele

D.R. Samarathunga[1], Y. Mathangasinghe[2], M. A. Jayawickrama[1], A. S. K. Banagala[1]
[1]National Hospital of Sri Lanka
[2]Faculty of Medicine, University of Colombo, Colombo, Sri Lanka

Keywords: Ewing sarcoma; kidney; varicocele; neoplasm

Introduction

Ewing's sarcoma is a primary malignant bone tumour of neuroectodermal origin accounting for the second commonest paediatric bone sarcoma (1). However, its involvement of the urinary tract as primary renal neoplasms is extremely rare, representing less than 1% of renal tumours (2). We present a case of primary Ewing's sarcoma of the kidney presenting with left-sided varicocele.

Case Report

A 24-year-old male presented with a left-sided scrotal lump of four years duration. Examination revealed a left-sided varicocele associated with an ill-defined mass in the left hypochondrium. On further questioning, the patient complained of left-sided intermittent flank pain, however, there were no significant urinary or systemic symptoms. Complete blood count, renal functions and erythrocyte sedimentation rate (ESR) were within normal ranges. Ultrasound scan showed a left-sided varicocele and a left-sided renal mass. Contrast-enhanced computed tomography (CECT) of the abdomen (Figure 1) revealed a left renal neoplasm compressing the vascular pedicle and para-aortic lymphadenopathy. The patient underwent left-sided radical nephrectomy. Left kidney, proximal ureter, left adrenal gland and an attached bowel mass containing distal ileum, caecum, large intestine and proximal sigmoid colon were removed during surgery.

Macroscopic examination revealed a mass measuring 150 x 95 x 90 mm mainly in the hilar region focally extending up to renal capsule and perinephric fat, replacing the normal renal parenchyma. Intestines were free of tumour invasion. Microscopy revealed a small round blue cell tumour with extensive necrotic area composed of solid sheets and nests of cells with hyperchromatic rounded to oval nuclei with scanty cytoplasm (Figure 2). Frequent mitotic figures, pseudorosette formation and microvascular invasion were seen. Adrenal

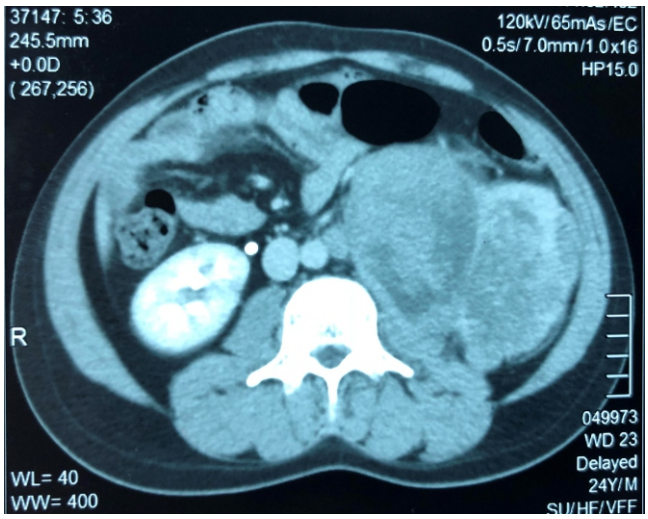

Figure 1. Contrast Enhanced Computed Tomogram (CECT) showing a left renal neoplasm compressing the vascular pedicle and para-aortic lymphadenopathy

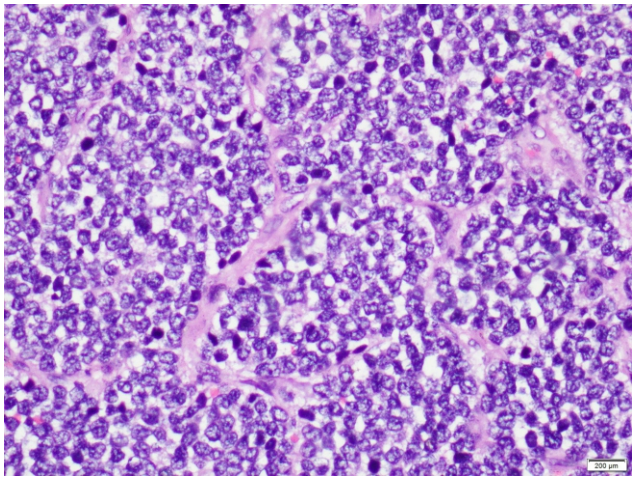

Figure 2. Haematoxylin and Eosin stain of the tumour showing sheets of small round blue cells with scant cytoplasm (40x magnification)

gland showed tumour emboli within vessels. Peri hilar region did not reveal lymph nodes. On immunohistochemistry, the tumour was negative for leukocyte common antigen (LCA), terminal deoxynucleotidyl transferase (TdT), pan-cytokeratin, WT1, Desmin, chromogranin and cluster differentiation 56 (CD56) excluding the possibility of a lymphoma and Wilms tumour. Immune-morphological features were of an Ewing's sarcoma (Figure 3). After surgery

Correspondence: Yasith Mathangasinghe
E-mail: yasithmathangasinghe@gmail.com
Received: 13-09-2019 Accepted: 27-12-2019
https://orcid.org/0000-0003-4641-5642
DOI: http://doi.org/10.4038/sljs.v37i4.8585

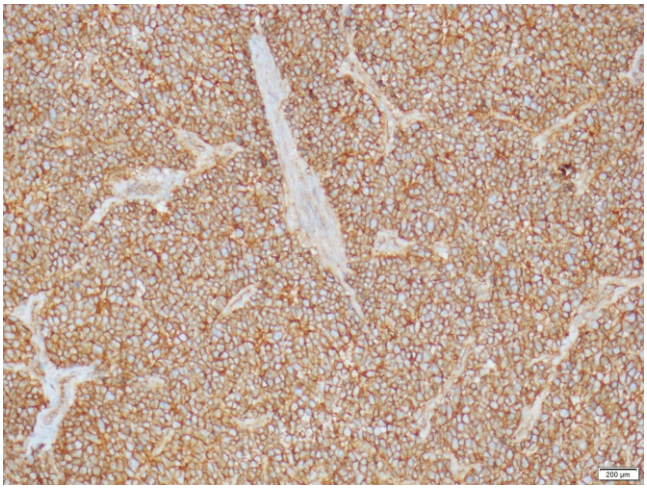

Figure 3. CD99 Immunohistochemical staining showing diffuse membrane positivity in tumour cells (20x magnification)

patient was referred to the National Cancer Institute for adjuvant chemoradiotherapy.

Discussion

Primary Ewing's sarcoma of the kidney is a rare entity associated with poor clinical outcomes (1). As in our case, it mainly affects young males at a median age of 28 years (1). Patients with primary renal Ewing's sarcomas commonly present with pain, haematuria and renal masses while a minority present with constitutional symptoms such as fatigue or loss of appetite (1, 2). In a case series, one-third of patients had metastasis at the time of diagnosis and 40% developed metastasis shortly after surgery (2). Commonest metastatic sites were lung (60%), liver (37%) abdominal lymph nodes (20%) and bones (16%) (2).

CECT may show large masses with necrotic, haemorrhagic and occasionally calcified areas but fails to provide any specific signs (3). Diagnosis is confirmed after histological and immunohistochemical analysis of surgical specimens (3). Microscopy comprises sheets of uniform small round cells with Homer-Wright rosettes positive for CD99 (4).

Ewing's Sarcoma is associated with translocations causing fusion of the EWS gene on 22q12 with a member of E26 Transformation Specific (ETS) family of transcription factors (1, 2). These fusion genes can be detected by fluorescent in-situ hybridization (FISH) techniques and are used as diagnostic markers in specialised centres.

Due to the rarity of this condition, specific treatment methods are not well established. Most of the cases are treated with radical nephrectomy with neoadjuvant and adjuvant chemoradiotherapy (5). Chemoradiotherapy has demonstrated significant survival benefits and improvement of symptoms even in patients with metastatic disease (1).

All authors disclose no conflict of interest. The study was conducted in accordance with the ethical standards of the relevant institutional or national ethics committee and the Helsinki Declaration of 1975, as revised in 2000.

References

1. Risi, E., et al., Clinical and pathological features of primary neuroectodermal tumor/Ewing sarcoma of the kidney. Urology, 2013. 82(2): p. 382-386. https://doi.org/10.1016/j.urology.2013.04.015
2. Parham, D.M., et al., Primary malignant neuroepithelial tumors of the kidney: a clinicopathologic analysis of 146 adult and pediatric cases from the National Wilms' Tumor Study Group Pathology Center. Am J Surg Pathol, 2001? 25(2): p. 133-46. https://doi.org/10.1097/00000478-200102000-00001
3. Ellinger, J., et al., Primitive neuroectodermal tumor: rare, highly aggressive differential diagnosis in urologic malignancies. Urology, 2006. 68(2): p. 257-62. https://doi.org/10.1016/j.urology.2006.02.037
4. Bing, Z et al., Primary Ewing sarcoma/primitive neuroectodermal tumor of the kidney. J Urol, 2009. 181(3): p. 1341-2. https://doi.org/10.1016/j.juro.2008.12.029
5. Celli R and G. Cai, Ewing sarcoma/primitive neuroectodermal tumor of the kidney: a rare and lethal entity. Archives of pathology & laboratory medicine, 2016. 140(3): p. 281-285. https://doi.org/10.5858/arpa.2014-0367-RS

Learning Points:

- Primary Ewing's Sarcoma of the kidney is a very rare entity.
- It commonly affects young males.
- Diagnosis is primarily by histological, immunohistochemical stains and genetic testing.
- Nephrectomy with neoadjuvant and adjuvant chemoradiotherapy is the preferred method of treatment.