

Extraosseous Renal Ewing Sarcoma: The “Blue” Tumour in kidney

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Abstract:

Introduction: Primary Ewing sarcoma (ES) is a distinctly rare tumour in the kidney and very limited cases were reported till date.

Case Presentation: Herein, we report a case of right renal ES in a young boy who presented with bleeding right renal tumour and underwent emergency right radical nephrectomy. Tumour cells are strongly positive for CD99 and FLI-1, thus confirming the diagnosis.

Conclusion: Therefore, this entity should always be one of the differential diagnosis while dealing renal masses in young population.

Keywords: Renal;extraosseus; Ewing’s sarcoma; blue tumour; CD-99, FLI-1

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Competing Interests: The authors have declared that no competing interests exist.

Consent: Consent was taken from the patient’s next of kin for publication of this case report.

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Introduction

Though ES is the second most common malignancy in bone and soft tissue in children and young adults, primary extraosseous ES is quite rare, indeed, just more than 100 cases of renal ES has been reported till date since first discovery in 1975 [1,2,3]. We report a case of right renal ES in a young boy who presented with bleeding right renal tumour and underwent emergency right radical nephrectomy in which the tumour cells are strongly positive for CD99 and FLI-1 [4,5].

Case report

A 19 years old Chinese boy presented with 2 weeks history of worsened right flank pain over a 2-month duration associated with low grade fever and abdominal distension. He denied any hematuria or recent trauma and other history was unremarkable. He was pale, tachycardic and there was right renal angle tenderness on examination. His computed tomography (CT) scan revealed a huge (16cm x 16cm x 12cm) heterogeneously enhanced right upper-mid pole kidney mass surrounded by diffuse perinephric hypodensities and streakiness in upper-middle pole, suggestive of intratumoural bleed with extension into right perinephric space. He then underwent emergency right nephrectomy. He had uneventful postoperative recovery other than minor partial wound dehiscence. He was well upon review in outpatient clinic 1 month postoperatively and referral made to Oncology team for further evaluation and possible adjuvant therapy.



Figure 1 (a) Axial view, (b) coronal view, CT showed large heterogenous mass occupying upper to mid pole of right kidney

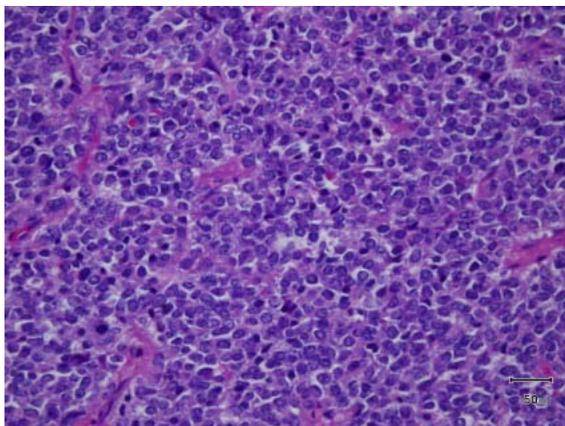


Figure 2 Microscopic photographs showing small blue round cells with almost uniform morphology in Ewing's sarcoma (H&E x400).

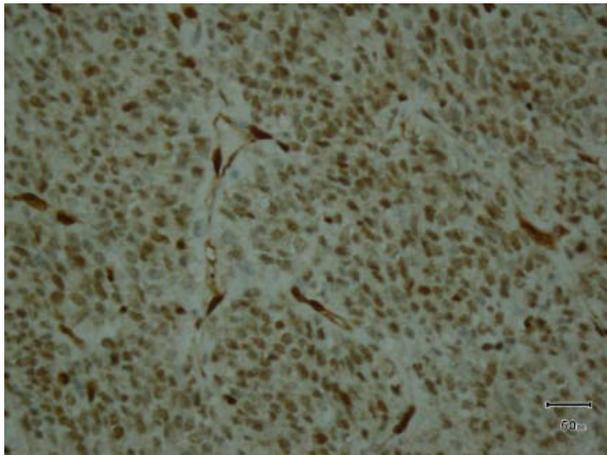


Figure 3 Microscopic photographs showing tumour cells are positive for FLI-1 immunohistochemical stain (x400).

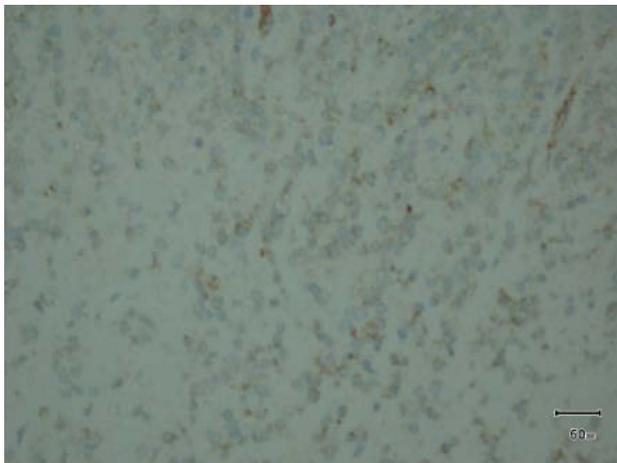


Figure 4 Microscopic photographs showing tumour cells are positive for membranous CD-99 immunohistochemical stain (x400).

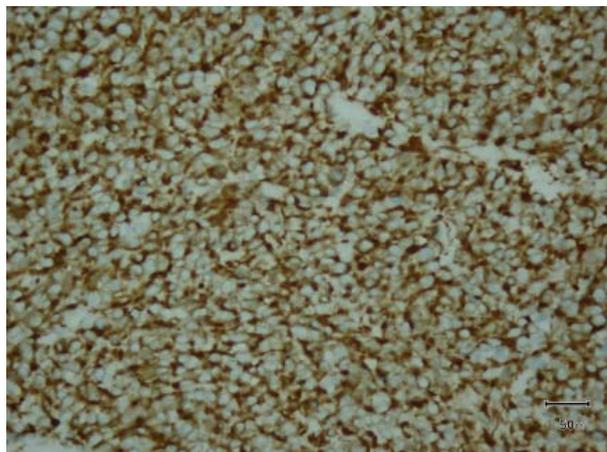


Figure 5 Microscopic photographs showing tumour cells are positive for vimentin immunohistochemical stain (x400).

Discussion

Renal ES is an aggressive neoplasm that predominantly affects male young adults [6,7,8,9]. Kidney sarcomas will usually remain silent until it grows beyond 5.5cm [7,9]. Clinical symptoms are indistinctive and the usually clinical findings are pain (85%), palpable mass (60%), and hematuria (37%) [8,9,10]. Radiological signs are non-specific and most of the imaging characteristics are indistinguishable from renal cell carcinoma (RCC) [6]. This “blue” tumour appears as ill-defined, large heterogenous mass with necrotic and hemorrhagic areas. On ultrasonographic examination it appears isoechogenic or hypoechogenic to the renal parenchyma, whereas on CT scan, the findings include areas of intratumoral hemorrhage or necrosis, peripheral hypervascularity, and diffuse calcification [7,8]. Some authors also described venous extension into the renal vein, inferior vena cava and right heart [8,11,12]. The differential diagnoses of renal ES are Wilms tumour, neuroblastoma, RCC, malignant lymphoma, metastatic renal involvement from sarcoma elsewhere in the body, and renal involvement by a primary retroperitoneal sarcoma [7,8]. Diagnosis of renal ES is very much rely on pathological findings and immunohistochemical staining. More than 90% of the tumours are typically positive for CD99 and FLI-1. CD99 is also positive in synovial sarcoma, however it will also show 90% positivity for pancytokeratin which was negative in our case [8,13]. Prognosis of disease is poor, with a 5-year survival of 60-70% in localised disease and 20-30% in metastatic disease. Common sites of metastasis include lung, liver, abdominal lymph nodes, and bone [8,14]. Treatment of primary renal ES includes surgery, chemotherapy and radiotherapy. As primary renal ES is an “once a blue moon” disease, till date, there are no standardised treatment protocol for it though these patients are generally treated with regimens for other type of ES. Surprisingly, the available literature does suggest a high response rate to chemotherapy, especially in local disease [14,15].

Conclusion

Primary renal ES is an unusually rare entity with poor prognosis that predominantly affects male young adults. Histopathological examination and immunohistochemistry are crucial to ascertain the final diagnosis before tailoring a treatment plan.

Consent

Patient was well informed that reporting rare disease is crucial in development of medicine and he was consented before the writing of this case report.

Conflicts of interest

Not applicable.

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