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Case Report

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Primary Ewing Sarcoma of the kidney: Case Report and Review of the Literature

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Abstract

The objective of this review is to discuss the features and nature of primary renal Ewing sarcoma, including incidence, presentation and management. We present our case of Ewing sarcoma of kidney along with imaging and pathological analysis. We also discuss the available literature and the current status of management.

Introduction

Primitive Neuroectodermal Tumors (PNETs) are small, round-cell tumors of neural crest origin typically found in the Central Nervous System (CNS) but more recently described in the periphery [1]. Peripherally located PNETs (pPNETs) are members of the Ewing's Sarcoma Family of Tumors (EFTs). It is the 2nd most common primary bone tumor in childhood. Less frequently it occurs in soft tissues [1].

Ewing's Sarcoma/Primitive Neuroectodermal Tumor (ES/PNET) is an extraordinarily rare primary tumor in the kidney. It was first described in 1975 by Seemayer and colleagues [2]. Only few cases of primary renal Ewing's sarcoma have been reported in the literature to date [1-10]. Renal Ewing sarcoma (RES) presents highly malignant, grows rapidly, and metastases early to the lung, bone and lymph node [11].

Case report

An 18 year old male presented to us with right flank pain over the past few days. There was no history of vomiting, haematuria or any bowel symptoms. Routine blood investigations were within normal limits.

A Contrast Enhanced CT (CECT) Abdomen and pelvis was done which revealed a mass in the Right Renal Fossa arising from the upper and mid pole of right kidney measuring 12.7 x 13.4 x 17.3 cm with heterogenous areas of necrosis. There was no renal vein or IVC thrombosis. Tumour was superiorly displacing and abutting the right lobe of liver and displacing IVC and renal vessels. It was not seen to invade surroundings organs. Para-caval and retrocaval nodes were enlarged, largest being 11 x 17 mm (Figure 1). A CT guided biopsy of the mass was done which revealed the mass to be a small round blue cell tumor which wasCD99 & FL11 positive and TLE1, CL7, EMA, WT1, Desmin negative suggesting it was an Ewing's sarcoma.

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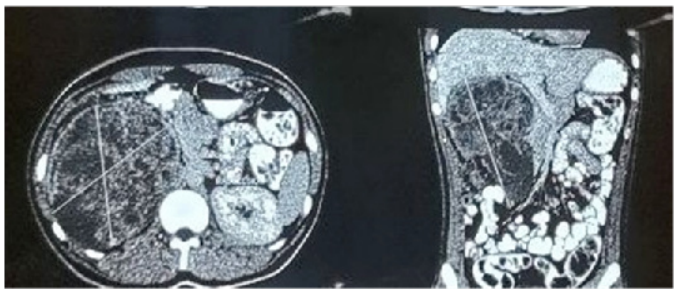

Figure 1: Preop CT images showing the extent of the mass.

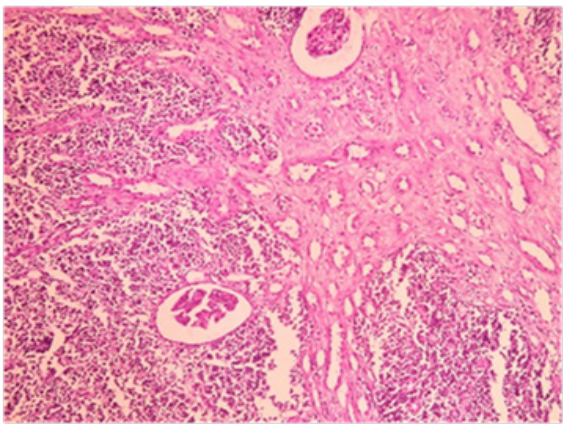

Figure 2: Small Round Cells in sheets with normal glomeruli & tubules.

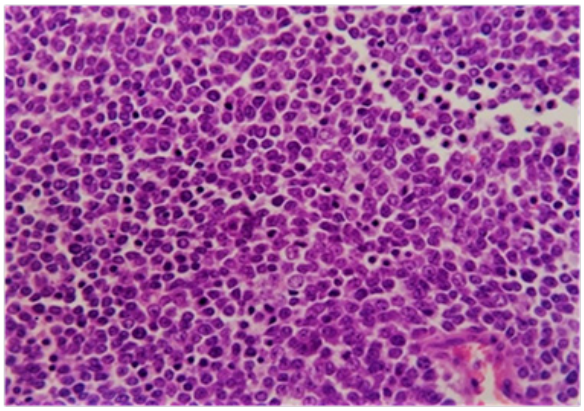

Figure 3: Small round cells in sheets suggesting ewings sarcoma (100x).

A right radical nephrectomy was performed and the mass was well encapsulated, abutting the liver and right colon, not invading any of these. Post operative course was uneventful. Tumor size was 19.5 x 15.5 x 15 cm occupying almost entire kidney with compressed renal parenchyma at periphery. It had a variegated appearance with friable brown areas along with occasional areas of haemorrhage and necrosis. Renal sinus was also involved with extension upto proximal ureter. All margins were free and adequate. Histopathology report confirmed the preop diagnosis of Ewing's sarcoma with the tumour being CD99, FLI1, CK7, PAX8 positive and BCL2 weak positive and the tumour as staged as PT1 as per CAP soft tissue sarcoma for visceral organ protocol (Figures 2-4). Patient received adjuvant chemotherapy and is fine on 1year follow up.

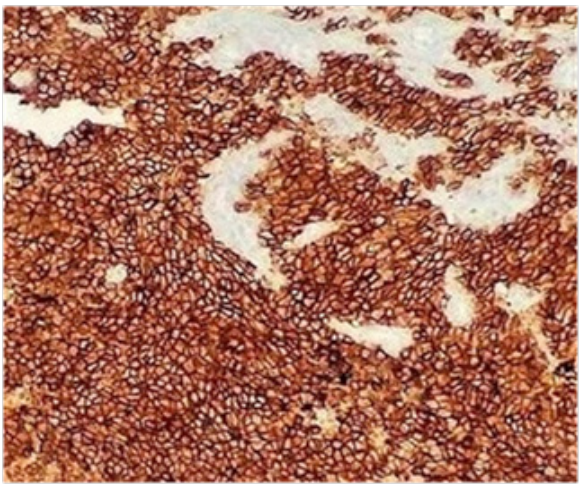

Figure 4: Immunopositivity for CD99.

Discussion

The entity peripheral PNET was first recognized by Arthur Purdy Stout in 1918 and is considered a member of the family of "small round cell tumours" [14]. The first report of rPNET(Renal PNET) was by Seemayer et al. in 1975 [2]. Primary renal localization is very rare. These tumors typically manifest in adolescents and young adults and have an aggressive behaviour. In a review by Kuroda et al., average age at diagnosis was 27.7 year [15] while that in Yuvaraja et al was 30 years [16]. Our case was diagnosed at an age of 18 years.

The disease usually has an aggressive course and the majority of the patients in are metastatic at presentation. Although against the norm, the largest Indian single centre series of 16 cases at Tata memorial had a majority (63%) of cases localized to the kidney.

Surgical excision remains the most important modality of management [12-14]. Despite aggressive treatment of these tumors by combination therapy with surgery, chemotherapy, and radiotherapy, the prognosis remains poor and overall 5-year survival rates have been reported at 45% to 55% and cases with advanced disease at presentation have a median relapse-free survival of only 2 years [1,11,13]. Radiation therapy is useful in treating these patients, especially when the R0 resection is not possible or residual disease is present. It is logical to give radiotherapy in the presence of Gerota's fascia involvement and positive surgical margins.

EWS/PNET of the kidney is typically characterized with a low enhanced large mass with multiple septum like structures and peripheral haemorrhage. In addition, a renal EWS/PNET is an aggressive and rapidly progressing tumor that is usually detected in adolescents and young adults with the presence of venous thrombosis and distant metastasis. When the adolescents and young adults have a large renal mass with such characteristics, EWS/PNET should be considered a diagnostic possibility.

Conclusions

PNET of the kidney is a distinct clinical entity with aggressive behaviour. This tumor should be considered in the differential diagnosis of renal tumors in adolescents and young adults. The tumor diagnosis is based on a classical histologic and IHC features com-

plemented by cytogenetics and molecular analysis. We conclude that aggressive multimodality treatment is recommended to manage these tumors.

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