CASE REPORT

Renal Rhabdomyosarcoma, Pleomorhic Variant in a 61-year old Male

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Renal rhabdhomyosarcoma is a rare subtype of renal sarcoma in adults that arise from skeletal muscle progenitor cells. Primary rhabdomyosarcomas are usually seen in the pediatric population and extremely rare in adults. This is a case of 61 year old, male who presented with a 3 week history of a palpable left upper quadrant mass accompanied by early satiety, anorexia, weight loss and dysuria. Ultrasound of whole abdomen revealed a hydronephrotic left kidney with no noted lithiasis and unremarkable right kidney. CT scan of the abdomen was chronic cystic pyelonephritis vs renal neoplasm, left paraaortic or perirenal lymphadenopathy suggestive of regional node metastasis. The case was diagnosed as Left Renal New Growth with consideration of Renal Cell Carcinoma (T2bN1M0). Hence the patient underwent cytoreductive nephrectomy. The histopathologic report revealed pleomorphic rhabdomyosarcoma. Literature review of renal rhabdomyosarcoma regarding symptomatology, diagnostic modality, treatment standard, adjuvant therapy and prognosis was done.

Key words: Rhabdomyosarcoma, pleomorphic, renal sarcoma, embryonal rhabdomyosarcoma

Introduction

Renal sarcomas are rare representing only 1-3% of all renal malignant tumors in adults. Rhabdomyosarcoma is the most common soft tissue sarcoma in children constituting more than 50% of case while it is rare and highly aggressive tumor in the adult population.[1,3] Primary renal sarcoma is highly malignant and prognosis is poor although early diagnosis and radical nephrectomy can prolong the patient's life.[2] Rhabdomyosarcoma is classified into four types; embryonal, spindle cell/sclerosing, alveolar and pleomorphic.[9] It has limited literature because of its extreme rarity.

The Case

Presented is a case of a 61-year old male, married, born again Christian from Taguig City who works as an overseas Filipino worker in Saudi Arabia with a palpable abdominal left upper quadrant mass. Three weeks prior to admission, patient noted a palpable abdominal left upper quadrant mass with an associated early satiety, anorexia, weight loss and dysuria. Consultation was done and on physical examination there was a movable non-tender mass on the left upper quadrant area measuring 15x12cm. Whole abdominal ultrasound revealed hydronephrotic left kidney with no noted lithiasis and a normal right kidney. On

whole abdominal CT scan the consideration was chronic cystic pyelonephritis vs renal neoplasm [Figures 1 & 2].

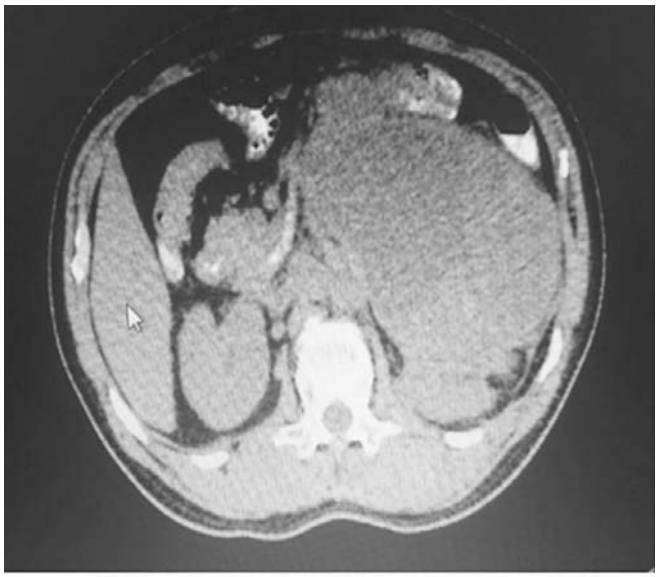

Figure 1. Plain abdominal CTscan.

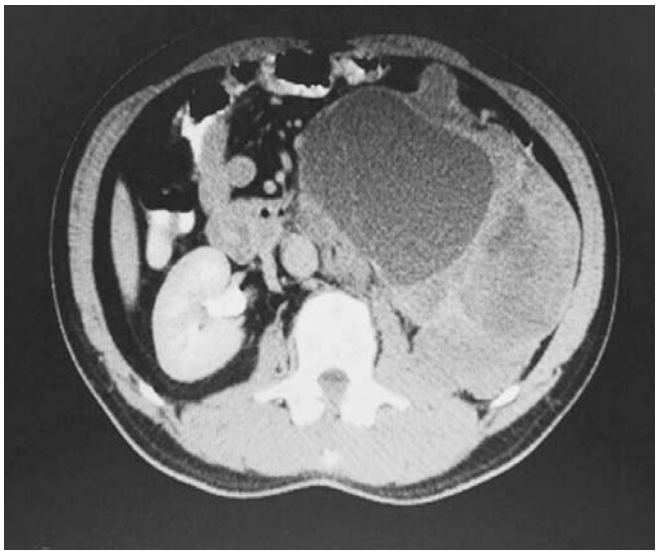

Figure 2. Abdominal CTscan with contrast.

Glomerular filtration rate was 0ml/min and 31.1ml/min for the left and right kidney, respectively. The patient was classifed as T2bN1M0 based on TNM staging. The patient underwent cytoreductive nephrectomy, left [Figure 3]. Intraoperatively, there was spread of tumor extending beyond the Gerota's fascia, and part of the tumor near the hilum was adherent to the aorta and inferior vena cava, the pancreas was displaced anteriorly by the tumor, while the mesocolon of the sigmoid was infiltrated by the tumor. The patient tolerated the procedure well and was subsequently discharged improved.

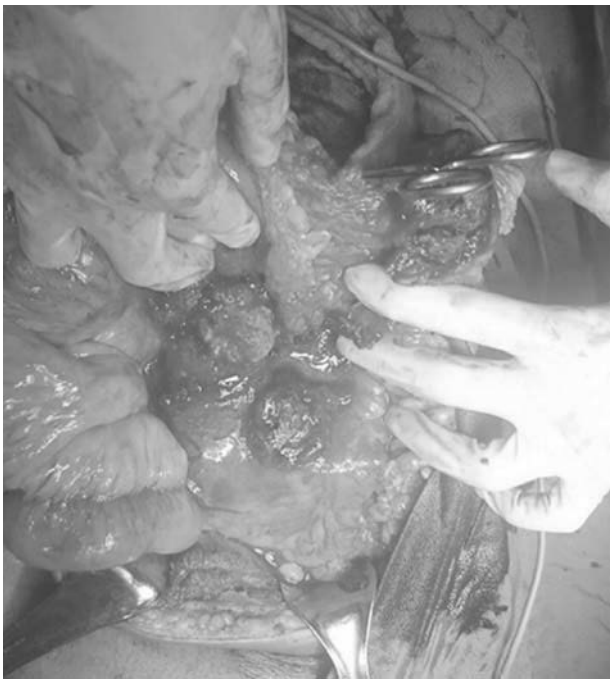

Figure 3. Intra-operative.

The gross specimen submitted showed a tan-brown, irregulary shaped, rubbery tissue with thin-capsule which measured 17.5cm x 13.0cm x 13.0cm and weighing 1,130grams [Figure 4]. Cut section shows tan-white, smooth solid surface with areas of hemorrhage measuring 17.0cm x 12.0cm x 9.0cm. The renal pelvis consists of tan-gray, smooth surface measuring 5.0cm x 4.0cm. There is no grossly identifiable renal cortex, medulla and ureter. Isolated lymph nodes consist of tan-brown, irregularly shaped, rubbery tissues with largest lymph node measuring 1.2cm x 1.0cm x 0.5cm.

The histopathologic examination of the mass showed pleomorphic tumor of elongated fusiform cells with cigar-shaped nuclei and prominent nucleoli. The tumor cells have

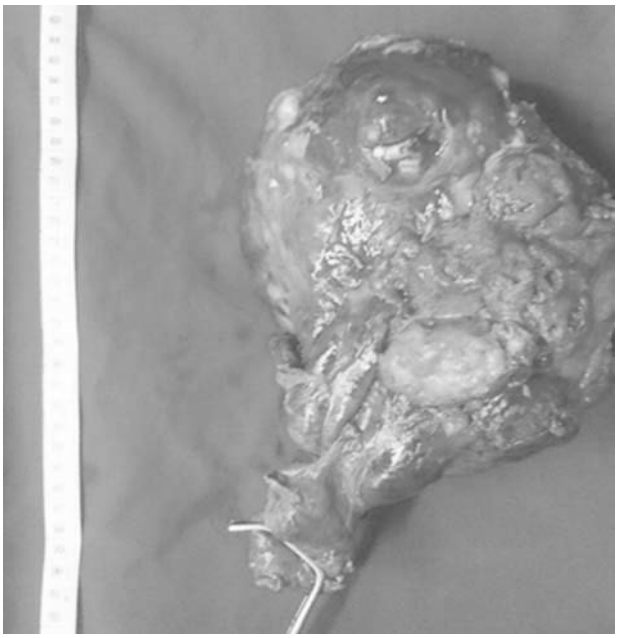

Figure 4. Specimen (Left kidney).

eosinophilic fibrillar cytoplasm with distinct cellular borders and giant cells. The spindle cells are separated by collagen fibers arranged in a storiform or whorled growth pattern. The lymphovascular spaces are positive for tumor invasion. The renal capsule, perirenal fat and renal vessels are positive for tumor invasion.

Discussion

Renal sarcoma is a rare case in adults and accounts for 1% of all primary renal malignancies.[4] Leiomyosarcoma is the most common histologic subtype of renal sarcoma accounting for 50-60% of renal sarcomas. Rhabdomyosarcoma is the least frequently reported. Adult cases are uncommon and arise mainly in large skeletal muscle.[4] Rhabdomyosarcoma is divided into four subtypes: embryonal, spindle cell/sclerosing, alveolar and pleomorphic.[9] Among the subtypes, embryonal rhabdomyosarcoma is the most common and has the best prognosis.[4] Genitourinary tract rhabdomyosarcoma arise most commonly in the urinary bladder, prostate, vagina and uterus.[3]

Primary renal sarcoma has the same clinical symptomatology with advanced renal cell carcinoma. The first and most common symptom is an abdominal mass.[5] The criteria for diagnosis of primary renal sarcoma include three components as defined by Grignon, et al.[6] Firstly, there should be no evidence of another sarcoma to exclude metastatic tumor in the differential. Secondly, a sarcomatoid renal cell carcinoma must be excluded. Finally, extension of a retroperitoneal sarcoma with secondary renal invasion can be excluded on histology. Median age of presentation for renal sarcomas is 49 years and the average size at diagnosis varies from 5.5 to 23 centimetres.[14]

Most renal sarcomas are indistinguishable from renal cell carcinoma on imaging and present as large soft-tissue mass with poor contrast enhancement. Biopsy of the specimen after resection can confirm the diagnosis. Myogenin and MyoD1, myogenic regulatory proteins expressed early in skeletal muscle differentiation are considered sensitive and specific markers for rhabdomyosarcoma.[4]

Treatment approaches to rhandomyosarcoma include surgery, radiation therapy and chemotherapy. However, radical nephrectomy is the gold standard for treatment of this kind of tumor[4] followed by adjuvant chemotherapy with vincristine, dactinomycin, and cyclophosphamide (VAC). Radiation therapy may be utilized for residual tumor and localized recurrences.[7,8]

Rhabdomyosarcoma of the kidney in the adult population is unusual, and only sporadic cases have been reported. This is a very aggressive tumor with dismal prognosis.[10] The most important predictors of outcome in patients with adult rhabdomyosarcoma are patient's age, tumor size, extent of disease, and margin status after resection. All histologic subtypes of rhabdomyosarcoma are aggressive malignancies with poor disease specific survival despite aggressive multimodality management.[11] The five-year survival rate is 82% in patients with retroperitoneal sarcoma, 73% in patients with sarcomas of the bladder, 44% in patients with prostate sarcoma and 39% in patients with sarcomas of the kidney.[12]

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