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Case Report

Giant renal tumor in adolescent: a diagnostic dilemma and a surgical challenge

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ABSTRACT

Giant renal tumors filling the entire abdominal cavity are rare in adolescents and pose a surgical challenge. Unlike their pediatric counterparts, where upfront chemotherapy can be administered without histologic confirmation, management of a giant renal tumour in adolescents follows that of the adults and is primarily surgical. Herein, we report a case of 17 years old male who presented with a 15×15 cm, 3.75 kg metastatic right renal tumour filling the entire abdominal cavity and displacing the great vessels and was managed by surgical resection, as multiple attempts of obtaining a percutaneous histologic diagnosis were unsuccessful.

Keywords: Giant renal tumour, Adolescent, Wilms tumour

INTRODUCTION

Renal tumors in adolescents are rare and account for 0.7% of all the cancers affecting the adolescents, with an age-specific incidence rate of 1.4 per million.[1]

Giant renal tumours filling the abdominal cavity are seldom reported in adults and rarely in children, but have not been reported in the adolescents. In sharp contrast to the management in children, where upfront chemotherapy for down staging can be an option, the management of a giant renal tumour in an adolescent follows that of adults and is primarily upfront surgical resection.

Herein, we report a case of 17 years old boy who presented with a 25 cm right renal tumour filling the entire abdominal cavity, with multiple pulmonary metastasis managed with upfront surgical resection as the preoperative percutaneous biopsies were inconclusive. The final histology was wilms tumor and the specimen weighed 3.75 kg and is one of the largest to be reported in adolescents.

CASE REPORT

A 17-years boy presented with complaints of right flank pain of 7 months duration and a mass in the right flank for 4 months, which was rapidly progressing in size. The abdominal examination revealed a 15×15cm lump filling the entire right half of the abdominal cavity and extending beyond the midline, associated with right non-reducing varicocele. The contrast enhanced computed tomography (CECT) scan of the abdomen and pelvis revealed a 18×12 cm mass involving the right kidney compressing the vena cava, abutting the aorta and filling almost the entire abdominal cavity (Figure 1). The CECT scan of the chest revealed multiple small pulmonary nodules suggestive of metastasis. A radiological diagnosis of metastatic renal tumour was made and the differentials considered were renal cell carcinoma, wilms tumor and primitive neuro-ectodermal tumor. In view of the large size, close relation with great vessels and presence of metastasis, the patient along with his parents were counselled and they chose percutaneous biopsy followed by systemic therapy in the hope of downstaging the tumor. The patient underwent

ultrasonography guided percutaneous biopsy and multiple cores were taken but the histopathology revealed only necrosis. The procedure was repeated 2 more times, using a multi-quadrant and multiple core technique, however the biopsy showed only necrosis. During this period the tumour grew significantly in size and thus they were counselled for open radical nephrectomy with a consent of only taking an adequate biopsy if found unresectable. A pre-operative embolisation of right renal artery was planned but was deferred by the radiologist in view of deranged coagulation parameters. The patient underwent right open radical nephrectomy via extended subcostal incision and an attempt was made to ligate the right renal artery in the inter-aortocaval region, however, because of the distortion of the anatomy and the presence of the mass in this region, it was not possible.

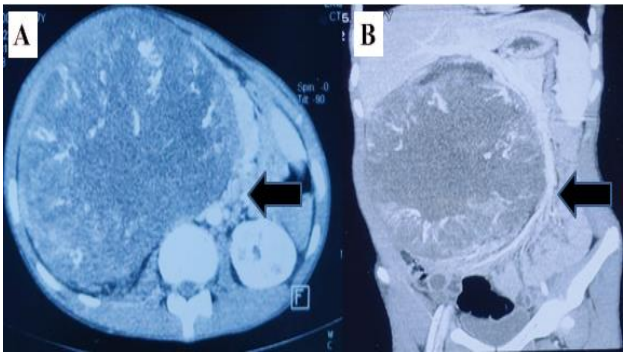

Figure 1: (A) and (B) CECT whole abdomen and pelvis - axial and coronal section depicting a large right kidney mass measuring 18×12 cm compressing the vena cava and abutting the aorta (arrow).

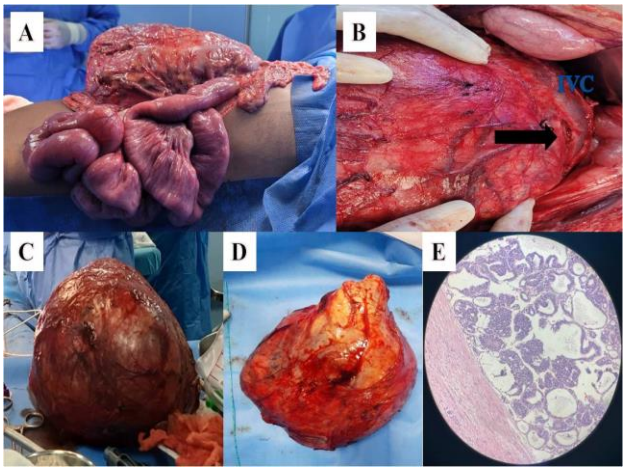

Figure 2: (A) Bulky renal tumour occupying entire abdominal cavity, (B) tumour dissected away from IVC with sharp and blunt dissection reaching the right renal vein (arrow), (C) and (D) bulky tumour sized 25×19×15 cm, and (E) micrography showing blastemal and epithelial component arranged in nodular and tubular pattern respectively (H&E, 400x).

After colon mobilisation, the mass was first mobilised inferiorly, laterally and superiorly and lifted off the great vessels to allow for identification of great vessels. The vena-cava was found to be compressed and stuck to the tumor and was freed by blunt and sharp dissection and the mass was removed along with the gerota's fascia covering and without any complications, rupture or spillage (Figure 2). The post-operative period was uneventful and patient was discharged on day 4.

On histopathological examination, the specimen weighed 3.75 kg and showed a tumour of size 25×19×15 cm. Microscopy showed a triphasic tumour comprising of blastemal, epithelial and stromal components with focal anaplasia (Figure 2). A final diagnosis of metastasic adult wilms tumour with triphasic pattern with focal anaplasia, stage IV was made. Patient was referred to medical oncology and adjuvant treatment was started as per high risk protocol for stage IV metastatic disease with weekly cycles of vincristine, actinomycin D and doxorubicin. At the last visit the patient has completed chemotherapy and has responded well and is under follow-up.

DISCUSSION

Occasionally, one may encounter renal tumours that fill almost the entire abdominal cavity and are deemed unresectable, however, such a scenario in an adolescent poses a diagnostic and surgical challenge. The commonest cause of renal tumour in an adolescent is translocation renal cell cancer, which typically presents as small mass with lymph node metastasis. On the other hand, wilms tumor and primitive neuro-ectodermal tumor, although rare, are more likely to present with large renal masses with involvement of the surrounding structures, and are likely to respond to pre-operative chemotherapy.[2,3] Thus, having a histologic diagnosis, prior to surgery can play a decisive role in managing large, difficult to resect renal tumors in adolescents.

Pre-operative chemotherapy has been shown to be highly effective in patients with wilms tumors and a significant size reduction can be expected in the majority of the patients.[3] Upfront chemotherapy, without a histologic diagnosis, can be administered in pediatric renal tumors, however, this does not hold true for adolescents. Ultrasonography guided percutaneous biopsy has established itself as a standard modality to obtain a histologic diagnosis and can be used to guide therapy.[4] However, we were unable to obtain a histologic diagnosis despite several attempts, probably because of the presence of large areas of necrosis. Also, in patients with wilms tumor, needle biopsy has been shown to cause biopsy site deposits and traditionally upstages the disease into "tumor spillage" and may also be associated with higher risk of local recurrence.[4] We also noted peritoneal deposits at all the sites of needle biopsy that required resection during surgery.

Our case is unique as the tumor weighed 3.75 kg, which is probably the largest in the adolescents, especially when considering intact removal.[5] Pre-operative percutaneous biopsy and angio-embolisation or ligating the artery in the interaorto-caval region, can ease the surgical process, but were not possible and the tumor had to be mobilised prior to hilar control, contrary to the oncological principles, to complete radical nephrectomy.

CONCLUSION

Giant renal tumours are rare in adolescents and obtaining a percutaneous biopsy can guide further treatment. Rarely, the biopsy may be non-contributory and an upfront surgical resection may be required.

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