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Case report

All paediatric renal masses are not Wilms' tumor!

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ABSTRACT

Background: Wilms' tumor being the commonest primary malignant renal tumor in childhood, makes Surgeons rarely think of other aetiologies. Renal cell carcinoma (RCC) in paediatric age group is a rare entity and most children with renal tumor are diagnosed unexpectedly following nephrectomy for presumed Wilms' tumor. Case presentation: We report an 8-year-old boy presenting with lump in the abdomen and haematuria for past 5 years. Abdominal ultrasound revealed a left renal mass measuring 10 × 10 cms, and a clinical diagnosis of Wilms' tumor was made. Radical nephrectomy was performed, following which histological diagnosis of clear cell variant of renal cell carcinoma was made. Conclusion: The possibility of a paediatric renal cell carcinoma should be borne in mind when a patient presents with pain in the flank and a renal mass. RCC in a child is often a histological surprise and needs documentation in the literature.

1. Introduction

The incidence of renal cell carcinoma (RCC) in childhood is estimated to be from 0.1% to 0.3% of all neoplasms and from 1.8% to 6.3% of all malignant renal tumors[1].Quite often diagnosis is revealed post operatively as a histological surprise to the treating Surgeons. The unfamiliarity of managing such cases in low volume centres often creates apprehension to both the patient care givers and Surgeons. Thus we make an effort to share our experience at a low volume tertiary care centre with the medical community in managing such cases.

Case report

An 8 year-old male presented with haematuria, dull aching pain and a lump in the left side of the abdomen. The lump was gradually increasing in size for the last 5 years, for which they were advised to undergo operation for suspected renal tumor. The general examination of the patient was non-contributory, as was tthe examination of respiratory, neurological & cardiovascular systems. Abdominal examination revealed a non-tender mass palpable in the left lumbarregion. Routine laboratory investigations including full blood count, chest X-ray and renal function s were normal. Urine examination showed plenty of RBCs in high power field.

Ultrasound examination of the abdomen revealed an echogenic mass in the upper part of the left kidney measuring 10 × 10 cm. No calcified areas were noted; Computerized tomography (CT) of the abdomen and pelvis revealed involvement of left renal pelvis and ureter but not renal vein (Figure 1). No signs of metastases visualized on imaging thorax, bones. A provisional diagnosis of Wilms' tumor tumor was made and left radical nephrectomy performed.

Grossly, the nephrectomy specimenweighed 400 grams and showed a solid spherical tumor with bosselated surface, measuring10 × 8 x 8 cm^3. The cut surface (Figure 2) showed multiple whitish nodular areas of varying size with a largest measuring 8 x 8 x 6 cm^3. They had central areas of hemorrhage and necrosis. Hilar lymphnodes were not identified. Normal kidney tissue could not be identified, however ureter showed infiltration by the growth. The renal capsule was intact. Microscopically, the tumor showed solid sheets of tumor cells, compartmentalized into solid acinar structure separated by vascularised fibrous septae (figure 3). Majority of tumor cells were large with eccentric nuclei, conspicuous nucleoli and irregular nuclear membrane and had clear cytoplasm (Figure 4). Foci of eosinophilic cells with moderate granular eosinophilic cytoplasm exhibiting pseudo-papillary growth pattern were also noted. The tumor margins were free of malignant cells.

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A final diagnosis of clear cell variant of RCC with Fuhrman grade 2 and clinical stage 1 was made. Adjuvant treatment options were discussed with patient care givers and three monthly follow up with serial ultrasound and chest x ray, urine for malignant cytology was undertaken. The patient remains in complete remission after 12 months of follow up.

Figure 1: Contrast enhanced computerized tomography (CECT) abdomen showing heterogeneously enhanced left kidney with intact capsule

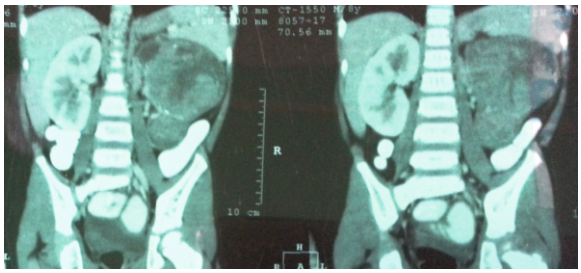


Figure 2: Cut surface of left kidney showing areas of haemorrhage and necrosis

Figure 3: Microscopic low power field showing Clear cell variant of RCC

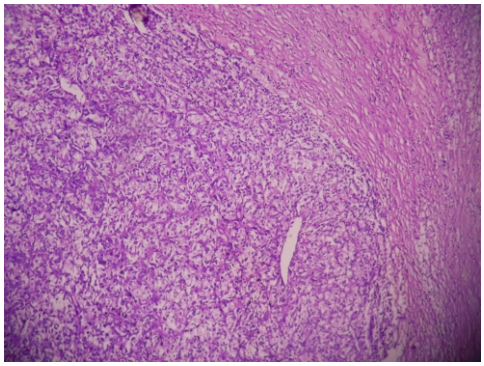
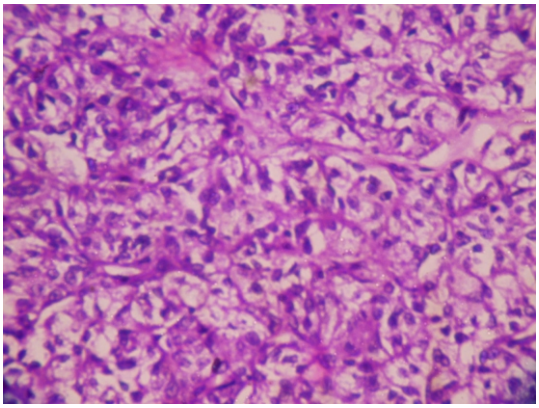


Figure 4: Microscopic high power resolution showing large eccentric nuclei with conspicuous nucleoli



Discussion

RCC is a rare disease in children and adolescents [2] and approximately only 160 well-documented cases have been published in the literature[3]. Though RCC is one of the differential diagnoses, practically most surgeons do not suspect it in a child. Indolfi et al [4], have noted male to female ratio of 2: 3 in contrast to other studies which showed 2:1 male predominance. Our patient was an 8 year old male child. Mean age of presentation of RCC is 10 years; in contrast Wilms' tumor is usually seen in less than 5 year olds [4].

The classical triad of Wilms' tumor, namely palpable mass, flank pain and haematuria, was noticed in our patient who presented with left sided lump in the abdomen. The literature documents 60% involvement of left kidney with 21% having palpable mass, 29.2% gross hematuria and 42.5% with pain abdomen[4]. Indolfi et al[4],also noticed that only 23.1% had a lump more than 10 cm. Our case also had a lump 10 x 10 cms showing ignorance and reluctance for radical surgery at such young age despite medical advice. Para-neoplastic phenomenon which is a well known entity in the adult RCC has been reported in few paediatric case series as well[5]. However our patient did not have such a phenomenon. Syndromic association of adult RCC with von Hippel-Lindau syndrome and Tuberous sclerosis is often reported; however such reports in paediatric RCC is scarce [4]. We did not perform chromosomal analysis in our patient due to lack of facility at our institute.

Fuhrman grading system is capable of predicting cancer specific survival independent of pathological stage [6]. Also Qayyum T et al [7], has demonstrated that combining grades 1 and 2 improves the prognostic ability of the Fuhrman grading system and a three tiered system combining grades 1 and 2 whilst keeping grades 3 and 4 separate is an independent predictor of survival. Our patient had grade 2 on histology.

Patients with tumor localized in the kidney have a good prognosis compared with patients with regional lymph node involvement or distant metastases [5]. Indolfi et al opines that no

optimal therapy has been defined for children with RCC. Surgery is the mainstay of the treatment and results in cure when the tumor is localized and completely resected. The roles of radiotherapy and immunotherapy are not clear. Pathologic staging [8] was performed according to the modified Robson staging classification system: stage I, localized disease confined by the renal capsule; stage II, localized disease invading renal capsule but confined by Gerota's fascia; stage IIIA, involvement of renal vein or inferior vena cava; stage IIIB, regional lymph node involvement; and stage IV, metastatic disease. The overall prognosis of children seems similar to that of adult patients and worsens as tumor stage increases [5].

No correlation was noted comparing surgical procedure (simple vs radical nephrectomy) with survival rate [4]. We performed radical nephrectomy in our case as we had preoperative diagnosis of Wilms' tumor with ureter involvement. Localised stage (stage I and II) have the best prognosis [9]. Indolfi et al [7], also had similar results in localized stage with most patients doing well in follow up. No effective chemotherapy has been reported in non-localized or relapsing disease [10].Minimal activity in stage III has been noted in spite of different chemotherapeutic regimes tried at various institutes[4].

Paediatric RCC was considered non responsive to radiotherapy (RT) and there is lack of consensus about its potential benefit in large trials. Castellano's et al [6], have noted favourable outcome in patients with lymph node involvement when RT was administered. Indolfi et al [7], in their small group of patients with stage IIIB noted increase in disease free survival. We did not give RT to our patient as the authors were of the opinion that close monitoring would be sufficient in such cases where long history of renal lump and absence of metastasis at the time of operating existed. MacArthur CA [10], has achieved complete response to recombinant IL-2 in child with metastases at presentation. However such promising results have not been used in practicality due to lack of randomised trials. Lack of funding/ insurance also makes such experimental trials impractical in a government set up like ours.

Metastases at presentation have been noted quite often in adult RCC but rarely in paediatric age group [9]. We also did not notice any metastases in the present case. A previous study on RCC in 84 children reported an actuarial survival rate of 60% at 2 years and 56% at 5 years [5]. Recurence and death have been noted after 15 months of diagnosis in stage 3 and 4 [4].

Conclusion

RCC in children should be considered when renal tumor is presented after 5 years of age. Considering the unfavourable outcome of RCC which is unearthed after surgery, careful monitoring is necessary. Chemotherapy, radiotherapy and immunotherapy, though promising, need to be established before considering them in protocol.

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