An Unusual Case of Clear Cell Sarcoma in Adult

Medical Science

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ABSTRACT

Clear cell sarcoma of the kidney is an uncommon renal neoplasm of childhood. It represents one of the most unfavorable tumors included in National Wilms' Tumor Study Group clinical protocols. Clear cell renal sarcoma is a rare tumor and comprises 4% of primary pediatric malignant renal tumors. It is known as an aggressive tumor with poor prognosis. We came across this rare tumor in an adult female (21 years-old). The case report is followed by discussion, stating the differentiating features between Wilms' and clear cell sarcoma, histological details, treatment, and prognosis. Clinically and radiographically, it resembles Wilms' tumor.

INTRODUCTION

Non- Wilms' renal tumors account for less than 10% of primary renal neoplasm of childhood1. Clear cell sarcoma also known as "Bone metastasizing tumor of childhood" by Marsden et al2 comprise 4% of all primary childhood renal tumours. It has a peak incidence during 3-5 years, with a male:female ratio of 2:1. CCSK is extremely rare in infants younger than 6 months and in young adults. Here we are reporting a 21 years old woman with clear cell sarcoma of right kidney who was managed with radical nephrectomy and chemoradiotherapy.

CASE PRESENTATION

A 21 years old woman presented with complaints of right sided abdominal swelling with flank pain. There was no history of vomiting, loss of weight or haematuria. On examination, the patient had pallor and a systolic blood pressure of 84 mmHg. The abdomen was distended and a firm, non-tender swelling was found occupying the right hypochondrial and lumbar regions. Blood and urine investigations including 24 hours vanillyl mandelic acid and chest radiograph were within normal limits. Abdominal computerised tomography showed a large heterogenous necrotic mass of size 121x150x180mm originating from lower pole of kidney.

Figure-1

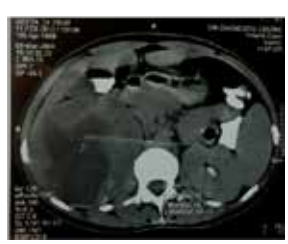

Figure-2

Fine needle aspiration cytology suggested Wilm's tumour or Germ cell tumour. Intravenous urography revealed minimally functioning right kidney with normal left kidney. Bone scan was negative. A right total nephrectomy was done through an intraperitoneal approach with an anterior subcostal incision. The macroscopic nephrectomy specimen revealed a well circumscribed tumour.

Figure-3

Formalin preserved specimen was sent for histopathological diagnosis which suggested clear cell sarcoma of kidney with focal microscopic penetration of renal capsule but negative resection margins consistent with stage II disease. There was a classic pattern of clear cell sarcoma of kidney consisting of cells with fine nuclear chromatin, pale cytoplasm and indistinct cell borders forming nest. No lymph node metastasis was present. Bone scan was negative

Concomitant chemoradiotherapy was performed. Adjuvant radiotherapy (4950cGy) was given. As adjuvant chemotherapy, Actinomycin (1.2mg/m2) with Vincristine (2mg/m2) and Doxorubicin (30mg/m2) with Vincristine (2mg/m2) were administered sequentially for one day every 3 weeks during one year period. No side effects were observed related to therapy. There was no evidence of recurrence or metastasis during the subsequent twelve months after therapy.

DISCUSSION

CCSK is an extremely rare neoplasm in adults. Amin and colleagues reviewed four new and eight previous cases of CCSK occurring in adult patients3. These are unilateral and unicenteric tumours although Manchanda et al4 reported a bilateral CCSK. CCSK is frequently confused with Wilms' tumour in children. However Argani and colleagues reported the striking differences between clear cell sarcoma and Wilms' tumour5.

Grossly CCSK tumours are large with mucoid texture, foci of necrosis and cyst formation. Tumour kidney junction is sharp but at the periphery there is entrapment of individual nephrons or collecting duct characterising its infiltrative nature. Most of the tumours have "classic pattern" where the tumour appears monomorphous with cords or nests of 6 to 10 cells separated by

evenly dispersed small vascular septa5. Viswanathan et al described various histological patterns of CCSK6. A 17% incidence of bone metastases has been noted in CCSK tumour in National Wilms' tumour study. Other sites of metastases are brain, regional lymph nodes, lung, liver and soft tissue.

CCSK is a highly malignant neoplasm. Recently, Hiradfar et al reported a case of CCSK in a six years old girl with atriocaval tumour thrombus7. Optimal treatment of adult patients with CCSK still remains unclear. Bhayani et al8 and Benchekroun et al9 reported that surgery with combination chemotherapy was an effective approach although Rosso et al10 stated that CCSK is highly resistant to radiation as well as chemotherapy. Surgery alone does not reduce chances of metastases. Adjuvant chemotherapy with doxorubicin alone or in combination has shown promising results in other studies conducted by Ali et al and Argani et al5.

CONCLUSION

Hence we conclude that aggressive surgical approach along with chemoradiotherapy is very much effective than surgery alone for preventing local recurrence and distant metastases. Accurate diagnosis is very important and therapy must also include doxorubicin regardless of the staging of the disease.

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