Clear Cell Sarcoma of Kidney Metastasizing to Orbit

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Introduction

Clear cell sarcoma of the kidney (CCSK) is rare comprising 4% of primary renal tumours in children.1 Clinical presentation resembles that of Wilm's tumour with both presenting as an abdominal mass, pain, and gross haematuria.3 CCSK usually occurs as solitary renal mass in male children aged 3years or younger. 3, 5

CCSK commonly metastasizes to bone which is associated with poor prognosis (survival rate <30%). 3, 5, 6 The skeleton and skull are the most common sites of bone metastases (3, 7). Metastasis also occurs to regional lymph node, lung, liver and rarely the brain.4 Childhood kidney tumors seldom metastasize into the cranial cavity unless it is a special histological variant (4).

We report the first case of a clear cell sarcoma with bony metastasis to maxilla and frontal bone.

Case report

A 3 year old child presented to a local paediatrician with complaints of abdominal pain, fever, nausea, vomiting and haematuria. Examination showed an abdominal mass measuring 7.5x7.5cm in the left lumbar region which was presumed to be splenic in origin. A complete blood count, immunochromatographic test for malarial parasite were unremarkable. Ultrasound of the abdomen was suggestive of a large mass in left lumbar region. Computed tomography (CT) scan of abdomen showed a 10x8x10cm mass abutting the left kidney. The mass enhanced with contrast and showed a non enhancing core suggestive of necrosis. Secondary hydronephrotic changes were noted. Though the mass displaced the aorta and pancreas there was no infiltration to adjacent visceral organs. Multiple enlarged nodes were noted in the peri-aortic region. USG guided fine needle aspiration biopsy

Figure 1: metastasis to maxilla (left) and frontal bone(right)

(FNAB) showed clear cell sarcoma of kidney. Treatment included radical nephrectomy. Three months later the child developed bony metastasis to maxilla(5x3cm) and frontal bone (2x4 mm; Fig 1). Biopsy confirmed the lesions to be metastatic CCS. Unfortunately the parents did not consent for further treatment. Eight months after presentation the child is alive with metastasis.

Discussion

Amongst pediatric tumors Wilm's tumor is most common (85%) followed by mesoblastic nephroma (5%), clear cell sarcoma (4%), rhabdoid tumor (2%) and other rare tumors.1 Clear cell sarcoma was first recognized by Kidd in 1970 (4). It has got several names in the literature as bone metastasizing renal tumor of childhood, undifferentited sarcoma of kidney or sarcomatoid renal tumor of childhood.

Clear cell sarcoma being rare is the most frequently misdiagnosed renal tumor. It is often confused with wilm's tumor. Among all clear cell sarcoma only 5% tend to metastasize, but metastasis carries a genuine poor

prognosis (1). The disease has got a male predisposition (1).

The uniqueness of our case is that we report the first case to be metastasized to maxilla. Our review of literature did not reveal even a single case of metastasis of clear cell sarcoma metastasizing to extra orbital tissues and presenting as a mass below the eyelid.

Conclusion

The first and foremost thing is that any child diagnosed as a case of wilms tumor on USG should always be confirmed with a USG guided FNAC because of its resemblance with clear cell sarcoma. The treatment of choice is definitely nephroureterectomy but the story never ends over there. After nephroureterectomy the child should be called for regular follow up to rule out metastasis and if present, there should be early diagnosis and treatment of the metastatic lesions to improve the prognosis for the patient.

Again the reverse side of the coin is that if a child presents with mass in the extraorbital tissues near the eyelid, metastasis from kidney should always be ruled out.

Consent

Verbal consent has been taken from the parents for revealing the details of the patient.

Ethical approval

Not applicable.

References

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