

Rare case of blastemal predominant adult Wilms' tumor with skeletal metastasis case report and brief review of literature

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ABSTRACT

Wilms' tumor (nephroblastoma) is extremely rare in adults, skeletal metastasis being still rarer. The clinical course of adult Wilms' tumor is very aggressive. The present case is a rare blastemal predominant adult Wilms' tumor presenting with skeletal metastasis. We report a case of 19-year-old female presented with severe low backache and colicky left loin pain of 3 months and progressive weakness of 15 days duration. Magnetic resonance image (MRI) of lumbosacral spine was reported as spinal metastasis with right renal mass. The patient underwent right radical nephrectomy and the tumor was histopathologically confirmed as adult Wilms' tumor. In case of adult Wilms' tumor, distant metastasis may be the first presentation and this possibility should be considered when an adult patient presents with flank pain and a renal mass.

Key words: Adult Wilms' tumor, renal tumor, skeletal metastasis

INTRODUCTION

Wilms' tumor (nephroblastoma) is the most common renal tumor encountered in children. It is extremely rare in adults accounting for less than 1% of all renal neoplasm.^[1] The common sites of metastasis described in literature are lung, liver, and lymph nodes. In both pediatric and adult Wilms' tumor, skeletal metastasis is very rare.^[2] Adult Wilms' tumor may have a more aggressive clinical course and a higher tumor stage at the time of presentation compared to that in children.^[3] The stage and histopathology of the tumor are the most important prognostic indicators. We hereby discuss a rare blastemal predominant adult Wilms' tumor

presenting with skeletal metastasis with a short review of available literature along with differential diagnosis of other relevant small round cell tumors.

CASE REPORT

A 19-year-old female presented with severe low backache and colicky left loin pain of 3 months and progressive weakness of 15 days duration. The pain was radiating to the groin and left leg. There was neither any history of prior abdominal surgery nor any history of trauma. Further there was no complaint of hematuria, pyuria, or calculuria. Menstrual cycle was regular.

Per abdomen examination revealed tenderness in left loin and spine.

Routine laboratory investigations including a full blood count, chest X-ray, and renal function tests were normal. Magnetic resonance image (MRI) of lumbosacral spine was reported as spinal metastasis with right renal mass [Figure 1a and b]. Computed tomography (CT) guided aspiration of spinal lesion was performed. The cytology smears were cellular and showed mildly pleomorphic round to oval cells attempting microacinar and papillary structures. Cytologically possibilities of blastemal predominant adult Wilms' and Ewing's

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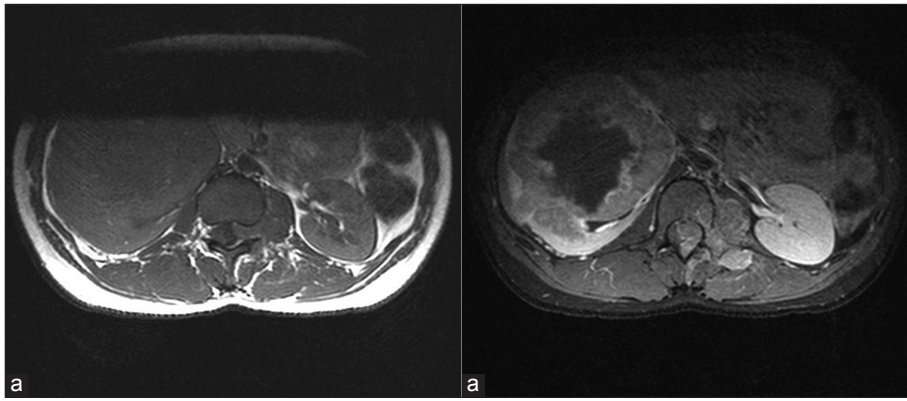


Figure 1: (a) Precontrast T1W axial images showing heterogeneously enhancing right renal mass and enhancing lesion in the body, left pedicle, transverse process and lamina of the L2 vertebral body with epidural component causing compression and displacement of thecal sac to right side. (b) Postcontrast T1W axial images of the same

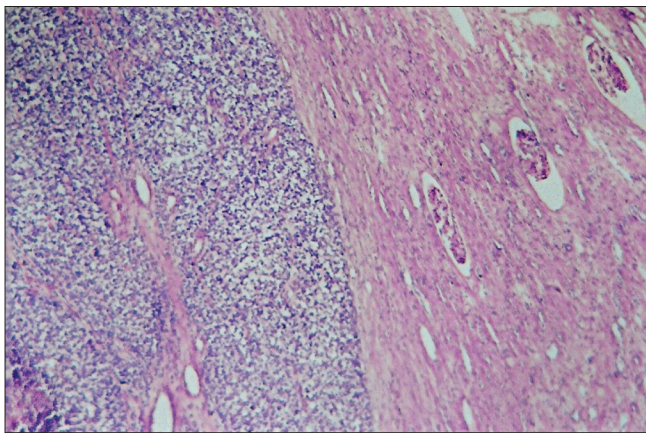


Figure 2: Monomorphic tumor cells with adjacent compressed normal renal parenchyma (H and E, $\times 20$)

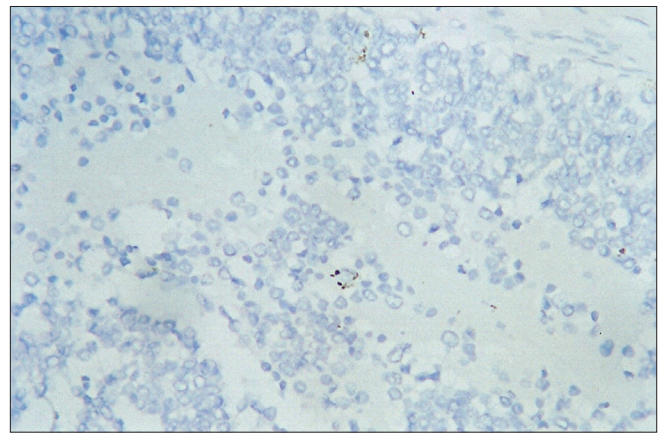


Figure 3: Immunohistochemically few tumor cells showing s-100 positivity

sarcoma/primitive neuroectodermal tumor (PNET) were considered.

The patient underwent right radical nephrectomy. The nephrectomy specimen showed a large tumor of $15 \times 10 \times 6$ cm with areas of hemorrhage and necrosis almost replacing the normal kidney.

Microscopically, the tumor comprised of monomorphous cells present as nests, islands, and sheets with intervening tumor necrosis and lymphoid collections. At areas, the tumor cells were seen around the blood vessels [Figure 2]. Ureter was free of tumor. The tumor cell on treatment with periodic acid schiff (PAS) agent did not show significant positivity, which favored the diagnosis of Wilms' tumor rather than Ewing's/PNET. Immunohistochemistry was performed with a panel of antibodies like vimentin, cytokeratin, CD45, synaptophysin, chromogranin, s-100, neuron-specific enolase (NSE), CD117, CD99. The tumor exhibited positivity for s-100, CD117, and NSE focally [Figure 3]. Rest of the markers was negative. Along with kidney lymph nodes around renal hilum and inferior vena cava were sent for histopathological examination. There was no evidence of metastasis in all 10 retrieved lymph nodes.

Further she received radiotherapy and six cycles of chemotherapy. Twelfth day after surgery, she was put on combined chemoradiotherapy. She received a total of 3060 cGy to spine over 17 fractions (180 cGy/day) and additional boost of 1080 cGy to tumor bed. She was offered pulsed intensive regimen with three drugs over 24 weeks. The chemotherapy schedule was dactinomycin $45 \mu\text{g}/\text{kg}/\text{day}$ at weeks 0, 6, 12, 18, and 24, doxorubicin $45 \text{mg}/\text{m}^2/\text{day}$ at third and ninth week and then $30 \text{mg}/\text{m}^2/\text{day}$ at weeks 15 and 21. Vincristine was given $1.5 \text{mg}/\text{m}^2/\text{day}$ at weeks 12, 15, 18, 21, and 24.

CT of the chest and abdomen at 3 months following surgery has not shown any evidence of recurrence of tumor or metastasis in other viscera. After completion of chemotherapy schedule, patient was lost to follow-up and presented with weakness of lower limbs and backache of 15 days duration one-and-half-year after surgical therapy. MRI of lumbosacral spine showed D11 and D12 extradural metastasis. There was no evidence of tumor bed recurrence. The patient underwent D10, D11, D12 laminectomy and excision of mass lesion histopathologically which exhibited similar morphology as that of the renal lesion.

There was no evidence of neurological improvement in the postoperative period and she was started on four drug regimen with inclusion of cyclophosphamide (J regimen of NWTSG-4), but patient refused to continue treatment after 4 weeks.

DISCUSSION

Wilms' tumor, named after the nineteenth century German surgeon Carl Max Wilhelm Wilms is a common malignant embryonic renal tumor of childhood but is extremely rare in people over 15 years of age. The histological appearance is characterized by marked structural diversity. Classic Wilms' tumor is composed of three types of cells undifferentiated blastemal, mesenchymal, and epithelial; although the occurrence of all three types in the same case is uncommon.^[4] Our case is a blastemal predominant Wilms' tumor.

Since there are no definite clinical data or radiographic investigations that can distinguish it from renal cell carcinoma, diagnosis is based on pathological evaluation. The diagnostic criteria necessary for adult Wilms' tumor suggested by Kilton *et al.* are primary renal neoplasm in age group of more than 15 years with histological features of embryonic glomerulotubular structure with immature spindle or round cell stroma and no areas of tumor diagnostic of renal cell carcinoma.^[5] The present tumor fulfilled most of the diagnostic criteria purposed by Kilton *et al.*

The relevant small round cell tumors, which should be considered in the differential diagnosis of this particular tumor, are Ewings/PNET and non-Hodgkin's lymphoma. The present tumor lacked the microscopical appearance of both these tumors and the corresponding immunohistochemical markers were also negative favoring the diagnosis of adult Wilms' tumor.

The prognosis of Wilms' tumor in adults is worse than in children because of the high recurrence, the lower response rate to chemotherapy regimens, and the advanced stage at the time of clinical presentation, like an asymptomatic abdominal mass in 75% of the cases. In spite of multimodal approach similar to childhood Wilms' tumor, the treatment outcome in adults is disappointing.

The majority of adult Wilms' tumor patients die of metastatic disease.^[6] Omachi *et al.* opined that prognosis of adult Wilms' tumor is very poor since many patients have unfavorable histology and no effective treatment guidelines have been established.^[7]

CONCLUSION

Adult Wilms' tumor is rare and diagnosed on histopathology mainly. These tumors behave more aggressively as compared to childhood tumors. The outcome is usually dismal in spite of multimodal treatment approach.

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