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Case Report

Paediatrics Section

Rhabdoid Tumour of Kidney with Metastasis to Ovary – A Rare Presentation

CHHANDA DAS, MADHUMITA MUKHOPADHYAY, ASHIS KUMAR SAHA, TAMANNYA PARVIN

ABSTRACT

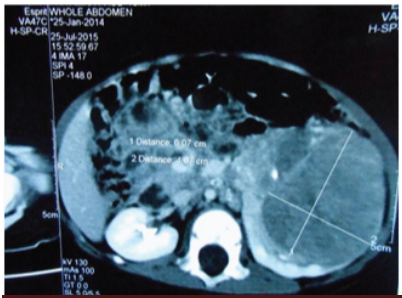
Rhabdoid tumour of kidney is a rare highly aggressive malignancy of early childhood. Here we are reporting an infant of six months who presented with mass abdomen in our hospital in the Department of Surgery. The Computerized Tomography finding was a hypo-echoic mass occupying most of the kidney. Laparotomy followed by nephrectomy was performed. The ovary also found involved by a tumour mass, which also was excised On gross appearance the mass was solid, nodular, and variegated measuring 6 cm x 5.5 cm occupying most of the kidney. Histopathologically, it was diagnosed as rhabdoid tumour of the kidney, ovary also showed metastatic deposits from kidney mass. It was reported as rhabdoid tumour of kidney metastasizing to ovary. Immunohistochemistry was found positive for vimentin in both kidney and ovary section. As it is a very aggressive tumour early diagnosis is essential for the treatment and patient survival.

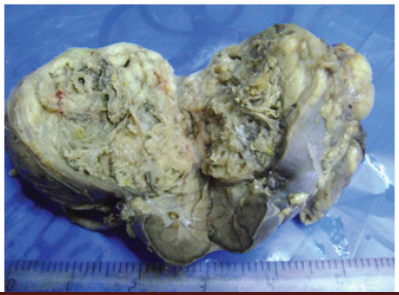
Keywords: Kidney tumour, Swelling, Vimentin

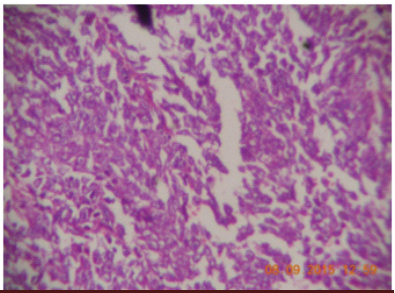
CASE REPORT

A 6-month-old female infant presented with gradually increasing swelling of abdomen for two months, in our hospital, at Surgery Department. The physical examination was most remarkable for a large intraabdominal mass. Clinically, it was suspected as kidney mass. Other systemic examination was normal. Routine investigations revealed no abnormality. Computed Tomography was done, it demonstrated a large heterogeneous mass located centrally within the kidney [Table/Fig-1]. Laparotomy followed by nephrectomy was done. The ovary also found involved by a tumour mass, which was also excised. The resected specimens of kidney and ovary were sent to our department for histopathological examination. Grossly, the kidney mass was solid, nodular and variegated in appearance, occupying most of the kidney, measuring 6 X 5.5 cm in size. In cross section, there were areas of haemorrhage and necrosis [Table/Fig-2]. On histopathological examination, the sections from kidney mass show diffuse and monotonous array of medium sized polygonal cells with spheroidal vesicular nuclei and large nucleoli, and intracytoplasmic eosinophilic hyaline globule [Table/Fig-3,4].

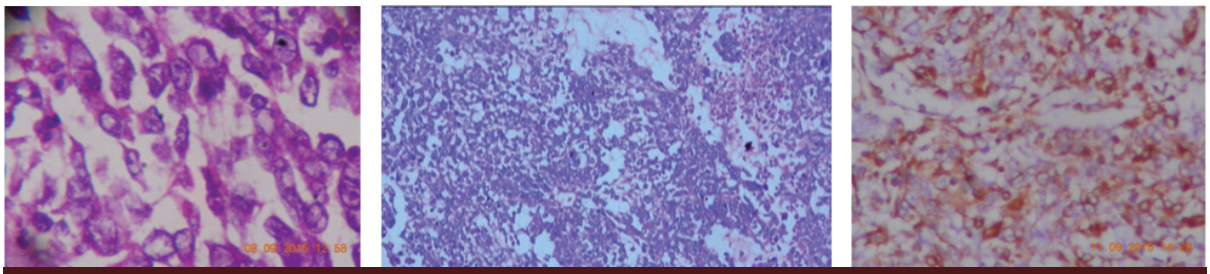
The histological section from ovary show metastatic deposits from kidney mass [Table/Fig-5].It was reported as rhabdoid tumour of kidney metastasizing to ovary. Immunohistochemistry study showed diffuse positivity for vimentin [Table/Fig-6]. So, it was confirmed as rhabdoid tumour of kidney. Ovary also showed positivity for vimentin [Table/Fig-7].



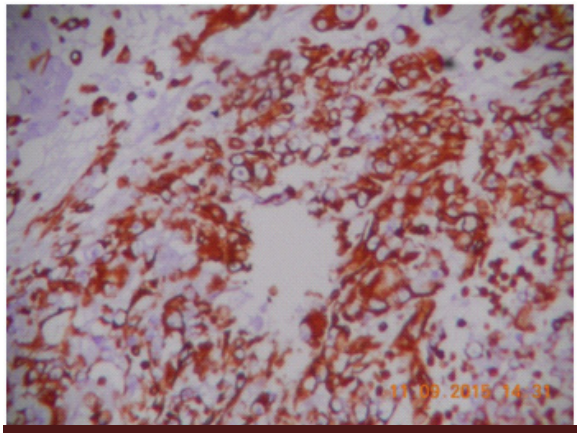




[Table/Fig-1]: CT scan demonstrated large heterogeneous mass within kidney.
[Table/Fig-2]: Gross nodular, variegated appearance of kidney.
[Table/Fig-3]: Photomicrograph showing sheets of tumor cells with vesicular chromatin, prominent nucleoli, (H & E, 100X). (images from left to right)



[Table/Fig-4]: Photomicrograph showing sheets of tumor cells with vesicular chromatin, prominent nucleoli (400x).
[Table/Fig-5]: Photomicrograph showing metastatic deposits in ovary (H & E, 100X).
[Table/Fig-6]: Immunohistochemistry staining of rhabdoid tumour kidney showing vimentin positivity (100x). (images from left to right)



[Table/Fig-7]: Immunohistochemistry staining of ovary showing vimentin positivity (100x).

DISCUSSION

Rhabdoid tumour of kidney is a rare highly aggressive malignancy of early childhood. Rhabdoid tumour usually metastasizes widely and causes death of the child within 12 months of diagnosis [1]. The most common sites of metastasis at presentation are the lungs, abdominal lymph nodes, liver, brain and bone. In our case ovarian metastasis found. Rhabdoid tumour comprises approximately 2% of paediatric renal malignancies [2]. The median age of diagnosis is 11 months, rare after three years and a predominance of males of 1.5:1 is seen [3]. Associations with embryonal tumours of central nervous system [4] and paraneoplastic hypercalcemia [5] reported. Rhabdoid tumours can occur sporadically or as part of hereditary cancer syndrome known as rhabdoid tumour predisposition syndrome [6].

Initially, it was described as rhabdomyosarcomatoid variant of Wilms tumour, but in 1981 Haas and colleagues used the term rhabdoid tumour because of the absence of muscular differentiation [7]. The histogenesis of rhabdoid tumour remains controversial, most likely an origin from renal medulla is suspected [8]. The gene mutated or deleted in malignant rhabdoid tumour of kidney is SMARCB1/ gene, also referred as SNF5 or INI-I or BAF47 [9]. Ultra structurally, the hyaline globule is made up of a tangle of intermediate filaments [7]. Immunohistochemically there is strong reactivity for vimentin and usually also for keratin but generally not muscle or neural markers. A wide variety of renal and extra rrenal tumours like Wilms tumour, congenital mesoblastic nephroma, renal cell carcinoma etc., may mimic rhabdoid tumour in H & E sections.

Chemotherapy is indicated as adjuvant treatment for rhabdoid tumour after primary tumour is surgically removed. Inspite of the recent intensified therapeutic regimes, the prognosis for rhabdoid tumour remain poor. Advanced stage, the presence of a CNS lesion and young age at diagnosis predict a dismal outcome [3]. In infants under six months at the age of diagnosis, survival was 8.8% in comparison with 41.1% survival for children with age at diagnosis of two years. Death usually within one year of diagnosis occurs in more than 75% of patients [10].

CONCLUSION

As it is a very aggressive tumour, early diagnosis is essential for treatment and patient survival. Although conventional microscopy is able to clarify most cases, immunohistochemistry may be required in some to show characteristic feature of rhabdoid kidney tumour. Resections to the extent possible, chemotherapy, and radiation therapy are often employed together in the treatment of this disease.

REFERENCES

[1] Reinhard H, Reinert J, Beier R et al. Rhabdoid tumours in children: prognostic factors in 70 patients diagnosed in Germany. *Oncol Rep*. 2008;19: 819-23.
[2] Lowe LH, Isuani BH, Heller RM et al. Pediatric renal masses: Wilms tumour and beyond. *Radiographics*. 2000;20(6): 1585-603.
[3] Tomlinson GE, Breslow NE, Dome J et al. Rhabdoid tumour of the kidney in the National Wilms' Tumour Study: age at diagnosis as a prognostic factor. *J Clin Oncol*. 2005;23: 7641-45.
[4] Bonnin JM, Rubinstein LJ, Palmer NF et al. The association of embryonal tumours originating in the kidney and in the brain, a report of seven cases. *Cancer*. 1984; 54:2137-46.
[5] Mayes L C , Kasselberg A G, Roloff J S et al. Hypercalcemia associated with immunoreactive parathyroid hormone in a malignant rhabdoid tumour of the kidney (rhabdoid Wilm's tumour). *Cancer*. 1984; 54: 882-84.
[6] Lee HY, Yoon CS, Sevenet N et al. Rhabdoid tumour of the kidney is a component of the rhabdoid predisposition syndrome. *Pediatr Dev Pathol*. 2002 ;5: 395-99.
[7] Haas JE, Palmer NF, Weinberg AG, Beckwith JB. Ultrastructure of malignant rhabdoid tumour of the kidney. A distinctive renal tumour of children. *Hum Pathol.* 1981. 12(7):646-57.

[8] Rosai J. Urinary tract In: Rosai and Ackerman's Surgical Pathology, 9th edition Mosby St. Louis. Missouri, 2004. p. 1250-51.
[9] Robert CW, Biegel JA. The role of SMARCBI/INI1 in the development of rhabdoid tumour. *Cancer Biol Ther*. 2009;8:412-16.
[10] Weeks D A, Beckwith J B, Mierau G W et al. Rhabdoid tumour of kidney, a report of 111 cases from the National Wilms' Tumour Study Pathology Center. *Am J Surg Pathol*. 1989;13: 439-58.

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