

Extrarenal Wilms' Tumor of the Ovary: A Case Report and Short Review of the Literature

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Abstract

Background: Wilms' tumor is a renal malignancy commonly occurring in childhood, with classic histologic features. Extrarenal Wilms' tumor is extremely rare, and only a few isolated cases have been described in the literature in the retroperitoneum, inguinal region, uterus, cervix, ovaries, skin, mediastinum, and chest wall. **Case:** A 14-month-old child presented with abdominal distention and a palpable mass in the right lower abdomen. Histologically, glomeruloid and tubular structures were seen within sheets of undifferentiated blastemal component. No teratomatous components were identified, despite extensive sampling, whereas a rim of ovarian tissue was present in one of the sections. **Results:** A diagnosis of primary ovarian Wilms' tumor was made. The patient was given chemotherapy and continues to be followed. **Conclusions:** Although very rare, Wilms' tumor may occur in sites other than the kidney and should also be considered as one of the differential diagnoses of an abdominal mass in children. Similar staging and treatment protocol to those for renal Wilms' tumor can be followed. (J GYNECOL SURG 28:306)

Introduction

WILMS' TUMOR IS THE MOST COMMON primary malignant tumor of the kidney in children. Extrarenal Wilms' tumor (ERWT) is exceedingly rare,¹ and varies in its clinical presentation depending upon its location and pressure effects on surrounding structures such as on the urinary bladder, bowel, ureter, blood vessels, bone, and nerves. As mentioned in the literature, the histologic characteristics and clinical course for ERWT are same as those of the renal counterpart; therefore, similar staging and treatment protocol can be followed.²

Case

A 14-month-old girl was admitted to the hospital with a history of abdominal pain and vomiting. There was no history of passing blood in stool or urine. Physical examination revealed a palpable mass measuring 12×10 cm in the right pelvic region. Findings from other systemic examinations were normal. Laboratory investigations were as follows: hemoglobin 10 g%, microcytic hypochromic picture and white blood cell count 6000/mm³. Urine analysis was normal and all biochemical investigations were within normal limits. Tumor markers (alpha-fetoprotein and beta human chorionic gonadotropin [β -hCG]) were within normal range. On ultrasonography, bilateral kidneys, liver, gallbladder, spleen, and pancreas

were normal. Contrast-enhanced computed tomography (CECT) scan was performed using non-ionic contrast medium, and it showed a heterogeneously enhancing lesion in the right pelvis and lower abdomen measuring 12×12cm. Both the kidneys were of normal size and density, and excreting concentrating contrast normally. Perirenal fascial planes were normal. Bilateral adrenal glands, liver, gallbladder, pancreas, and spleen were normal. No free fluid was seen, and a final diagnosis of heterogeneously enhancing lesion in the pelvis and lower abdomen of ovarian origin was given on CECT. The patient was taken for exploratory laparotomy. Perioperatively, a 12×12 cm lobulated intra-abdominal mass was identified arising from the ovary. It was not adherent to surrounding organs. Complete surgical excision of the mass was performed, in addition to removal of right ovary and tube.

Pathologic findings

Grossly, the excised tumor was binodular, measuring 10×8 cm with solid and cystic areas, and was associated with foci of hemorrhage and necrosis (Fig. 1). The ovary could not be seen separately. The fallopian tube was not involved. Microscopically, the tumor showed a mixture of undifferentiated blastema, and epithelial and mesenchymal tissue. The blastematosus areas were extremely cellular and composed of small

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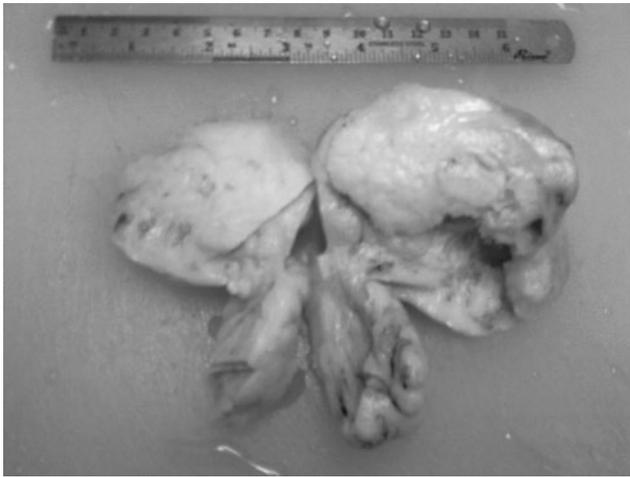


FIG. 1. Binodular tumor; cut section is gray-white with areas of hemorrhage and cystic degeneration.

round-to-oval cells admixed with primitive epithelial tubules and scattered glomeruloid structures. Blastema and epithelial elements were separated by stroma consisting of spindle cells of indeterminate type. There was no evidence of anaplasia. Teratomatous elements were not seen, despite exhaustive sampling. In only one of the sections was a peripheral rim of residual ovarian tissue seen (Fig. 2). Immunohistochemistry was performed. Tubules were positive for keratin and stromal cells were positive for vimentin (Fig. 3). A diagnosis of primary ovarian Wilms' tumor was made.

Postoperative period

The postoperative period was uneventful and patient was referred to pediatric oncology for further treatment. She received four cycles of chemotherapy, which included two

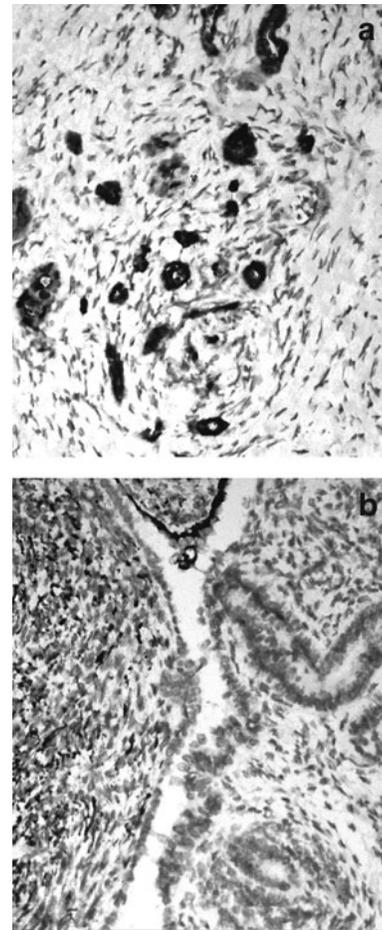


FIG. 3. Immunohistochemical (IHC) staining: Epithelial membrane antigen (EMA) positivity in epithelial component (a) and vimentin positivity in mesenchymal component (b) (IHC×200).

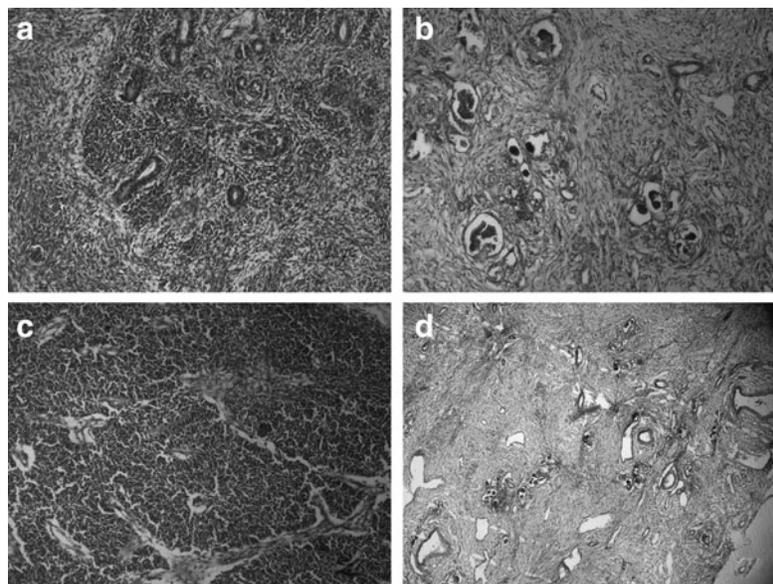


FIG. 2. Wilms' tumor showing characteristic triphasic histology, including epithelial (tubules and glomeruli), blastemal, and mesenchymal components (a–c) (hematoxylin and eosin [H&E]×100). One of the sections contains ovarion tissue (d) (H&E×40).

drugs, vincristine and actinomycin, and has been on regular follow up since then.

Discussion

Wilms' tumor is a malignant tumor with a characteristic histologic appearance, seen typically in children and almost exclusively involving the kidney. ERWT are exceedingly rare and may be seen in the inguinal region, endocervix, uterus, epididymis, ovary, testis, and any place in retroperitoneum along paravertebral area. The age at presentation usually ranges from 2 months to 10 years.³ Their histogenesis, morphology, clinical staging, behavior, prognosis, and response to therapy are similar to those of renal Wilms' tumor.

The origin of ERWT is controversial and unclear, as this condition is very rare. However there are different schools of thoughts including

1. Origin from metanephric blastema, as the majority of the tumors occur in the retroperitoneal region. However, the presence of ERWT cephalad to the kidney refutes this hypothesis.⁴
2. Origin from primitive mesodermal tissue; occurrence of ERWT in the cervix, vagina, and inguinal canal support this theory.⁵
3. Connheim's cell rest theory: according to this hypothesis, origin of tumor is from a cell with persistent embryonal potential to undergo malignant transformation at any point in time.⁶

Based on their probable histogenesis, ERWT can occur in two settings. First, those arising within teratomas; and second, those in which there is no evidence of a teratomatous origin.¹ Histogenesis of the second type is controversial.

While diagnosing a tumor as ERWT, extension from the intrarenal tumor should be ruled out and other differential diagnosis should also be considered. In this case, there was no evidence of primary renal tumor either by imaging or perioperatively. The most important differential diagnosis is immature teratoma composed largely of nephroblastic tissue. No teratomatous element was identified in this case, despite exhaustive sampling. The second possibility could be retiform Sertoli-Leydig cell tumor because of the presence of tubular structures. However, the presence of blastemal tissue and the absence of typical Sertoli-Leydig areas excluded this possibility. Adenocarcinoma and malignant mixed müllerian tumor should also be included as a differential diagnosis and they can be distinguished from Wilm's tumor by the presence of typical carcinomatous foci and the absence of blastemal and glomeruloid elements.

Staging and management of extrarenal nephroblastoma is the same as for its renal counterpart.⁷ All cases are treated by surgery with postoperative adjuvant chemotherapy. Radiotherapy

is reserved for unresectable tumors and those with distant metastasis. The presence of anaplasia indicates poor prognosis with increased resistance to chemotherapy.⁸ In this case, features of anaplasia (polyploidy and nuclear and mitotic atypia) were not seen. The patient was treated surgically with postoperative adjuvant chemotherapy and is on regular follow-up.

Conclusions

Although very rare, Wilms' tumor may occur in sites other than the kidney and should also be considered as one of the differential diagnoses of an abdominal mass in children. Morphology, behavior, prognosis, and response to therapy of this tumor are similar to those of renal Wilms' tumor; therefore, similar staging and treatment protocol can be followed.

Disclosure Statement

No competing financial interests exist.

References

1. Oner U, Tokar B, Acikalin MF, Ilhan H, Eskisehir NT. Wilms' tumor of the ovary: A case report. *J Pediatr Surg* 2002;37:127.
2. Waingankar VS, Meisheri IV, Patel MP, Ramesh S, Naregal A, Muthal P. Extrarenal Wilms' tumor: A case report and review of literature. *Indian J Cancer* 1995;32:135.
3. Ramachandra C, Attili VS, Dadhich HP, Kumari A, Appaji L. Extrarenal Wilms' tumor: A report of two cases and review of literature. *J Indian Assoc Pediatr Surg* 2007;12:145.
4. Broecker BH, Caldamone AA, McWilliams NB, Maurer H, Salzberg. A Primary extrarenal Wilms' tumor in children. *J Pediatr Surg* 1989;24:1283.
5. Fernandes ET, Kumar M, Douglas EC, Wilimas J, Parham DM, Rao BN. Extrarenal Wilms' tumor. *J Pediatr Surg* 1989; 24:483.
6. Waingankar VS, Meisheri IV, Patel MP, Ramesh S, Naregal A, Muthal P. Extrarenal Wilms' tumor: a case report and review of literature. *Indian J Cancer* 1995;32:135.
7. Teerthanath S. Extrarenal nephroblastoma. *J Lab Physicians* 2011;3:59.
8. Beckwith JB. New development in pathology of Wilm's tumor. *Cancer Invest* 1997;15:153.

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