Teratoid Wilms' tumor: a case report

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Teratoid Wilms' tumor is rarely seen and is a description used only recently. The term describes classical nephroma with a diversity of cell types and tissues. In this reported case, the epithelial component consisting of squamous areas made up 70 percent of the tumor; no criteria of dysplasia nor any nephroblastomatosis areas or endodermal elements were presented. Although it is reported that teratoid Wilms' tumor is not usually aggressive or metastatic, a case of unilateral teratoid Wilms' tumor in a 2.5-year-old-boy who died because of metastatic disease is presented and the literature reviewed.

Key words: pediatric "teratoid" Wilms' tumor, Wilms' tumor.

Wilms' tumor is the most common renal tumor of childhood. It is an embryonal neoplasm that is generally assumed to arise from metanephric blastema. Typically, it exhibits a triphasic histological pattern of blastema, epithelium and stroma[1-4]. In addition to these components, some small foci of heterologous elements may be encountered. The term teratoid Wilms' tumor was introduced by Variend et al.[5] to define a variant of nephroblastoma in which the heterologous tissue predominated. We report a child with unilateral Wilms' tumor which included teratoid as well as typical nephroblastic components.

Case Report

A 2.5-year-old boy was admitted to our clinic with hematuria and a right-sided mass noticed by his mother two years previously. The physical examination revealed a right-sided abdominal mass and right cryptorchidism. The laboratory findings were normal except for mild anemia and hematuria. Bone marrow aspiration was evaluated as normal. Vanillylmandelic acid was negative in 24-hour urine analysis. Ultrasonographic investigation showed anterior and inferior displacement of the right kidney and that the right echogenic mass was 10x15 cm. Intravenous urography showed a right-sided renal mass distorting the pelvicalyceal system, and hydronephrosis was present on the left kidney, resulting from extrinsic obstruction of the pelviureteric junction (PUJ). Computerized tomography (CT) confirmed that the mass arose from the right kidney, and it showed multiple periaortic lymph nodes. Chest x-ray was normal.

Exploratory laparotomy was performed. A firm well-encapsulated and lobulated renal tumor was found, completely excised, and a biopsy was taken from enlarged periaortic and mesenteric lymph nodes. Exploration of the contralateral kidney was evaluated as normal except for mild hydronephrosis. Hydronephrosis on the left kidney was assumed to be due to enlarged lymph nodes displacing and compressing the PUJ extrinsicly. The tumor was histopathologically diagnosed as teratoid Wilms' tumor.

The tumor was accepted as stage III because of periaortic and mesenteric lymph node metastases according to the National Wilms' Study Group 4. The child was given chemotherapy that included vincristine, dactinomycin, doxorubicin and whole abdomen irradiation (total 2,570 rad). Follow-up CT three months later revealed persistent multiple periaortic lymph nodes and a hydronephrotic left kidney. A second laparotomy was planned, but because of complications due to chemotherapy and irradiation, the operation was postponed. The patient was lost to follow-up and received no further chemotherapy. After an interval of six months, the patient was admitted with pulmonary

insufficiency. A computerized tomography examination of the abdomen and chest showed progressive hydronephrosis on the left kidney as well as pulmonary metastases. Although the chemotherapy protocol was changed, the patient died due to pulmonary and renal failure.

Pathology

Macroscopic Findings : The right residual normal kidney was compressed by a large, encapsulated tumor (13x13x9 cm). The kidney was seen as a band 3 cm in length under the capsule. The cut surface of the tumor was lobulated, pink, with scattered necrotic foci and lipomatous tissue. Some small cysts were also identified in the tumor. The renal artery and the ureter were grossly uninvolved.

Microscopic Findings : Microscopic examination of the resected specimen showed histology of classical triphasic Wilms' tumor which included blastematous, epithelial and stromal components. Solid nests of squamous epithelium were also present, some with central keratinization (Figs. 1 and 2). There was some mesenchymal differentiation showing the characteristics of striated muscle. The epithelial component consisting of squamous areas made up 70 percent of the tumor, and no criteria of dysplasia were identified. Although numerous samples were taken, there was no nephroblastomatosis area (Fig. 2). No endodermal elements were seen in the tumor tissue. There were metastases consisting of blastematous components in periaortic and mesenteric lymph nodes.

Fig. 1. A triphasic Wilms' tumor with solid nests of squamous epithelium (HEx40).

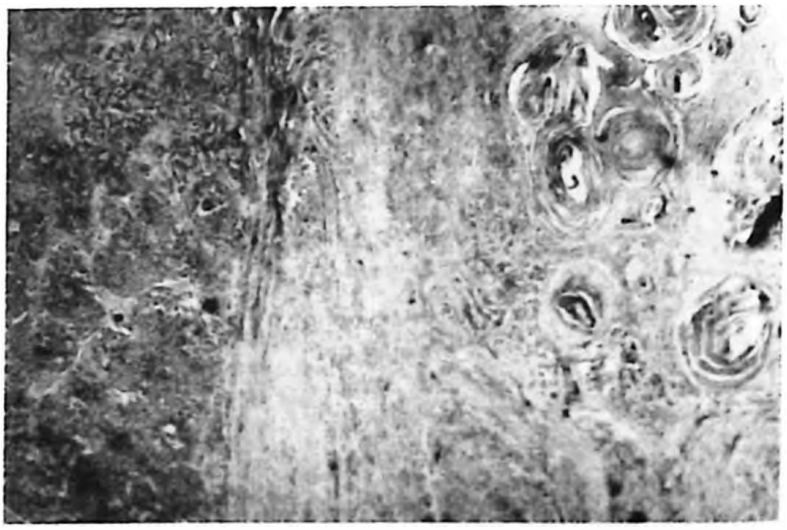

Fig. 2. High power view of Wilms' tumor with squamous nests (HEx100).

Discussion

Wilms' tumor is an embryonic tumor of mesodermal origin that contains blastematous, epithelial and stromal elements[6]. Embryonic tumors are totipotential in nature. Totipotency is variable and may depend on the stage of nephrogenesis at which tumor induction occurs[5,7]. Recently,. a rare variant of the typical triphasic Wilms' tumor, referred to as a teratoid Wilms' tumor, has been described, in which a more diverse differentiation causes a tumor that contains features of teratoma and nephroblastoma[5]. Although it is well known that nephroblastomas can include a diversity of cell types[2], in teratoid Wilms' tumor there is clear predominance of teratoid elements, comprising more than 50 percent of the tumor[3,5,8]. In our case, while the tumor histologically presented as classical triphasic Wilms' tumor, the epithelial component consisting of squamous areas made up 70 percent of the tumor. For this reason, it was accepted as a teratoid Wilms' tumor. Although many studies have described mucinous epithelium or an enteric-type epithelium including endodermal elements in the tumor tissue[9-11], no endodermal elements were seen in the tumor tissue of our case. Teratoid tumors of other organs, such as the intraocular teratoid medulloepithelioma, have also been described[12]. Ward et al.[13] reported a case of sacrococcygeal teratoma containing nephroblastoma in 1974. Teratoid Wilms' tumor differs from true intrarenal teratoma, the differential diagnostic criteria of which have been described by Beckwith[1] and Glazier[14]. According to them, epithelial and mesenchymal differentiation originated from the totipotent renal blastema, and "organogenesis" is the clue for differential diagnosis.

In our case, there was unilateral cryptorchidism as well. The presence of associated malformations (inguinal hernia, cryptorchidism, and club feet) suggests that there is a relationship between embryogenesis and carcinogenesis in this disease[8]. Among 290 patients admitted to St. Jude Children's Research Hospital with the diagnosis of Wilms' tumor, three of them were classified as teratoid Wilms' tumor. Their prominent clinical and histopathologic features were: 1) bilateral involvement, 2) tumor extension into renal pelvis causing ureteral obstruction, renal failure and hypertension, and 3) a close relationship between teratoid Wilms' tumor and the nephroblastomatosis complex. The well differentiated nature of the teratomatous elements may make this tumor somewhat resistant to chemotherapy and radiotherapy, depending on the extent of teratomatous involvement within the lesion[8]. In our case, there was unilateral involvement, and although there was no evidence of dysplasia, the patient died from metastatic disease. Hydronephrosis on the contralateral kidney in our case was thought to be due to enlarged lymph nodes around the PUJ.

To render optimal therapy for the teratoid type Wilms' tumor, consideration must be given to the clinical and histopathologic characteristics of the disease[8]. Inducing cellular modulations and nonepithelial differentiation of preoperative chemotherapy and radiotherapy have been reported, but the epithelial and rhabdomyoblastic components usually show no evidence of chemotherapeutic effect[15-17]. It is therefore not surprising that teratoid Wilms' tumor is resistant to chemotherapy. In the case reported by Kotiloğlu et al.[10], no regression in the volume of the tumor was observed after preoperative chemotherapy. In our case, neither chemotherapy nor radiotherapy was given preoperatively.

Teratoid Wilms' tumor is not usually aggressive or metastatic[3,4,5,8], if the proportion of blastemic elements in each tumor is considered. But, accompanying anaplastic elements may increase the likelihood of metastases and progression to death. Williams et al.[11] reported a case of teratoid Wilms' tumor who died because of metastatic disease, as in our case and it was reported that the major component of the tumor was adipose tissue. The interval time without chemotherapy and the postponed second look operation caused the bad prognosis in our case. In conclusion, teratoid Wilms' tumor is a rare variant of triphasic Wilms' tumor. Although it is reported that most of the teratoid Wilms' tumors are not usually aggressive or metastatic, our case died because of common metastatic disease without accompanying anaplastic elements. To our knowledge, our case is the twelfth case reported as teratoid Wilms' tumor and the seventh case reported as unilateral teratoid Wilms' tumor.

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