Extrarenal Teratoid Wilms' Tumor in Association with Horseshoe Kidney

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Abstract Extrarenal teratoid Wilms' tumor is a rare variant of Wilms' tumor. Literature shows only 17 reported cases. We present this right-sided teratoid Wilms' tumor in a 3-year-old boy for its rarity in association with the horseshoe kidney. The tumor was found in the anterior aspect of the right kidney and consisted of triphasic Wilms' tumor element along with the presence of heterologous components.

Keywords Extra renal teratoid Wilms' tumor · Complete excision · Wilms' tumor · Horseshoe kidney

Introduction

Teratoid Wilms' tumor is a rare histological variant of Wilms' tumor. Extrarenal teratoid Wilms' tumors have been reported in the retroperitoneum, sacrococcygeal region, testis, uterus, inguinal canal, and mediastinum [1–5]. The first case of teratoid Wilms' tumor was reported in 1984 by Variend et al. [6]. Till now, a few cases of extrarenal teratoid Wilms' tumors in association with the horseshoe kidney have been reported in the literature. In our case after complete excision of the tumor, the prognosis has been good without pre or postoperative chemotherapy or radiotherapy. Teratoid Wilms' tumor is a

rare histologic variant of the classical Wilms' tumor, containing predominantly heterologous tissues [7]. Regarding staging and management of extrarenal Wilms' tumor, the behavior of this tumor appears parallel to that of a similarly staged intrarenal Wilms' tumor. Hence, the staging and management protocols of intrarenal Wilms' tumor can be applied to an extrarenal location [8]. If the tumor is resectable and involves only one side of the horseshoe kidney, resection is recommended by the present National Wilms' Tumor Study Group (NWTSG). We present this unusual case of extrarenal teratoid Wilms' tumor in association with the horseshoe kidney so that pediatric clinicians as well as pathologists are made aware of the rare tumor.

Case History

A 3-year-old boy was admitted in our department with a mass of size 10 cm × 11 cm in the right loin of the abdomen present for 3 months with no history of fever or hematuria. He is not having any other congenital anomalies except the horseshoe kidney. He has normal blood pressure. The laboratory findings, including hematological, biochemical, and urine examinations, were within normal limits. Serum α -fetoprotein, measured by radioimmunoassay, was normal.

Clinical examination revealed a firm and nontender mass in the right loin. X-ray abdomen revealed a mass of soft tissue density, occupying the right side of the abdomen, displacing bowel loops. The ultrasound examination showed a large, solid, predominantly hyperechoic mass, measuring 10 cm in longest dimension. X-ray chest was normal (Fig. 1). Intravenous urography (IVU) revealed the right-sided nonexcreting mass in delayed film (Fig. 2).

Computed tomography (CT) of the abdomen showed a large, 10 cm × 9 cm size, cystic lesion with enhancing solid

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Fig. 1 Pre operative CT

areas with in it (Fig. 3). The left kidney appears malrotated. Both kidneys appear united by a band of tissue. Bowel loops were found to be displaced by the mass. The rest of the abdominal viscera were normal. There was no evidence of lymphadenopathy on CT examination. Impression was the horseshoe kidney with a mass lesion on the right side.

The child underwent exploratory laparotomy. Under general anesthesia, the abdomen was opened by a generous supraumbilical transverse incision. The lateral peritoneal reflection over the tumor was opened and the colon was reflected medially. There was no thrombus found in renal vessels and IVC. Vessels feeding the mass from renal vessels were ligated and divided. The mass of size about 10 cm×11 cm was identified in the anterior aspect of the right kidney (Figs. 4, 5 and 6). The mass was mobilized

Fig. 2 Pre operative X-ray chest

Fig. 3 Post operative CT

from the right kidney, ureter, isthmus and removed completely with its capsule without rupture. Both kidneys with capsules, band of tissue connecting both kidneys, both ureters, and pelvis were kept undisturbed. A single small right-sided iliac lymph node of size about 1/2 cm×3/4 cm was removed. Abdominal wound was closed with a drainage tube and removed on the fifth postoperative day. He was kept under broad-spectrum antibiotic for 14 days. The operation was successful. His post operative period was uneventful. One year after the surgery, he was reviewed with CT abdomen, X-ray Chest, IVU, and α -fetoprotein (AFP). All the results were normal.

Fig. 4 Per operative

Fig. 5 Per operative

Pathology

On gross examination showed the removed mass weighed 550 gm and the macroglobular cystic mass measured 10 cm×12 cm. The cut section showed uniloculated thin-walled cyst filled with thin mucoid fluid. One focus showed a polypoidal mass. Multiple sections were taken from the solid mass and cyst wall (Fig. 7).

Histopathological Examination Sections from the solid mass showed undifferentiated blastemal elements with attempted glomeruli and primitive tubules in a pseudorosette formation. Some places showed densely packed small solid

Fig. 6 Per operative

Fig. 7 Cross section

elements. The stroma showed focal collection of primitive and mature adipocytes, muscle cells, myxoid fibroblasts with spindle cells, blood vessels, and cystically dilated glands lined by flattened epithelial cells.

There were large areas of necrosis, hemorrhage and minimal calcification. The section from the cyst wall showed minimal fibrous thickening and was lined by flattened epithelial cells.

Special Stain Study Special stain with PAS showed PAS-positive secretion. Van Giesan stain showed muscular tissues in yellow colour and connective tissue in orange colour (Figs. 8, 9 and 10). Excised single node study showed reactive follicular hyperplasia.

Fig. 8 Microphoto shows small cells and blastemal elements with attempted glomeruli tubules and glomeruli formation

Fig. 9 Microphoto shows collection of primitive and mature adipocytes with myoid stroma

Discussion

As the tumor in our patient displayed mainly mature teratomatous elements comprising predominantly blastemal component with attempted tubules, abortive glomeruli, tubular structures with pseudorosettes, and immature cartilaginous elements, along with classical triphasic elements of Wilms' tumor and was extrarenal in location, the most appropriate diagnosis of extrarenal teratoid Wilms' tumor associated with the horseshoe kidney was made. The extrarenal location of teratoid Wilms' tumor has led to the debate on whether the origin is embryonic or neoplastic, but most authors believe the origin of this tumor to be the embryonic remnants of mesonephric tissue [9, 10].

Occurrence of extrarenal Wilms' tumor associated with the horseshoe kidney is very exceptional and the diagnosis is almost always made after surgical intervention. The tumor can be located in the retroperitoneum, uterus, cervix, testes, and skin and even in the thorax. The exact mechanism whereby a Wilms' tumor occurs in extrarenal tissues is not

Fig. 10 Micro photo. Undifferentiated blastemal elements in solid sheets

Fig. 11 Pre operative IVU

known [11], even if the histological characteristics are the same as in intrarenal Wilms' tumor.

Wilms' tumor is associated with congenital anomalies in 10–13 % of cases. Aniridia is present in 10 % children; hemihypertrophy is noted in 2–3 %. Other genitourinary malformations are present in 5 % of cases, mainly cryptorchidism, hypospadias, double collecting system or fused kidney. Rarely Wilms' tumor has been found in a horseshoe kidney [12]. Resistance to chemotherapy and radiotherapy has been reported due to the presence of mature heterologous compounds [13, 14].

Fig. 12 Post operative IVU

AFP is a useful diagnostic tool for hepatocellular carcinoma and germ cell tumors [15]. AFP is a common tumor marker in pediatric neoplasms, and increased serum levels are usually encountered in tumors arising from tissues producing AFP during fetal life. However, elevation of such protein is rarely found in patients with Wilms' tumor. A very few cases of Wilms' tumor associated with elevated serum AFP have also been reported [16]. A teratoid Wilms' tumor with elevated serum AFP level was reported in a 12-month-old boy presenting with a lumbar mass. Teratoid Wilms' tumor with raised serum AFP level is an unusual type of tumor [17]. The serum AFP level is elevated in 75 % of yolk sac tumors, 70 % of embryonal carcinomas, and 62 % of teratomas [18]. β -hCG was not done in this case.

Conclusion

Final diagnosis could only be arrived on histopathological examination. Apart from excision of the tumor, our patient was not put on radiotherapy or chemotherapy regimen. More than 1 year after the surgery, our patient was doing well, and close follow-up has been instituted in order to monitor any future recurrence or metastasis (Fig. 11). Although no further active treatment is indicated, long-term follow-up is advised to evaluate history, clinical examination, and repeated imaging scans. His computed tomography abdomen, IVU (Fig. 12), and AFP were normal after 1 year.

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