Teratoid Wilms’ tumor in a child: A report of a rare case

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Abstract
Teratoid Wilms’ tumor is an unusual variant of nephroblastoma in which heterologous tissue predominates. We report a case of teratoid Wilms’ tumor in a 2-year-old male. Right sided abdominal mass was the presenting complaint. Ultrasonography of the abdomen showed a mass in the right kidney. Histopathological examination revealed blastemal, epithelial, and mesenchymal components along with areas presenting heterologous elements. More than 75% predominance of squamous differentiation with the keratin pearl formation was observed. The patient underwent nephrectomy and was followed post-operatively for 1 year and was normal.

Key words: Nephroblastoma, squamous differentiation, teratoid Wilms’ tumor

Submission: 23-04-2012 Accepted: 19-12-2012

Introduction
Wilms’ tumor is the most common primary renal tumor of childhood and characterized by recognizable attempts to recapitulate different stages of nephrogenesis.[1] The term teratoid Wilms’ tumor is an unusual variant of nephroblastoma in which heterologous tissue predominates.[2] The classical triphasic combination of blastemal, stromal and epithelial cell types is observed in the vast majority of lesions, although, the percentage of each component is variable. Rarely, other heterologous elements are identified, including squamous or mucinous epithelium, smooth muscle, adipose tissue, cartilage, and osteoid and neurogenic tissue. Fewer than 30 cases of teratoid Wilms’ tumor have been reported until date.[3]

Case Report
A 2-year-old male with a complaint of right sided abdominal mass of 1 month duration revealed a palpable right abdominal mass on the physical examination. Ultrasonographic examination of abdomen presented a large, sharply demarcated predominantly hyperechoic mass of 7 cm × 5.5 cm × 4 cm in right para vertebral region in the location of normal kidney with areas of hypoechogenicity. No normal kidney tissue was identified on the right side. No calcification is noted. No intravascular echogenic focus was identified on color Doppler sonography. Left kidney was normal in position and echotexture. However, computed tomography scan was not performed as the patient was very poor to afford the test. Pre-operative routine investigations; serum urea, creatinine, and the blood urea nitrogen were normal. Serum alpha feto-protein levels were within normal limits. With a provisional diagnosis of Wilms’ tumor, right sided nephrectomy was carried out and the specimen was sent for the histopathological examination.

On gross examination, the tumor measured 6.5 cm × 5 cm × 4 cm and was weighing 250 g. It was well-circumscribed and encapsulated. Cut sections presented homogenous gray-white tumor tissue, soft, and granular in the texture. Ureter and renal veins were not involved by the tumor.

Microscopically multiple sections examined from different areas showed classic triphasic combination of blastemal, stromal, and epithelial cell types [Figure 1]. The tightly packed blue cells are the blastemal elements, immature tubule in the picture is the epithelial element and spindle

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| **Quick Response Code:**  |
| Website: www.ijabmr.org   |
| **DOI:** 10.4103/2229-516X.112248 |
cells are stromal elements. The heterologous elements are predominant and composed of squamous epithelium with abundant keratin pearl formation [Figures 2 and 3] and constitute about 75% of the area from where the sections were examined.

Considering the above histopathological features, a diagnosis of teratoid Wilms’ tumor was arrived. The patient was referred to the medical oncology department. No post-operative chemotherapy was advised to consider the diagnosis of teratoid Wilms’ tumor. Patient was doing well 1 year post-surgery. After that, the patient was not in contact for further follow-up.

**Discussion**

Wilms’ tumor constitutes the prototypical example of a neoplastic process that faithfully recapitulates embryogenesis at the morphologic and molecular level. It is seen primarily in infants with 50% of the cases occurring before the age of 3 years and 90% before the age of 6 years. The classic location for Wilms’ tumor is the kidney. Right and left kidneys are equally affected.

The term “teratoid Wilms’ tumor” was first used by Variend et al. in 1984. In 1988, Fernandes et al. proposed the criteria of more than 50% heterologous component for the diagnosis of teratoid Wilms’ tumor. Until date, fewer than 30 cases of teratoid Wilms’ tumor have been reported. About 38% of cases are bilateral. In the present case, the tumor was unilateral and the contralateral kidney was normal. Raised alpha feto-protein levels in occasional cases were reported. Alpha feto-protein levels were normal in the present case. Teratoid Wilms’ tumor is predominately a tumor of childhood and only one case occurring in an adult has been reported until now.

Behavior of teratoid Wilms’ tumor is usually not aggressive with a favorable outcome. Teratoid Wilms’ tumor is relatively resistant to chemo and radiotherapy and surgery is the treatment of choice. However, a few author recommend chemotherapy in these cases regardless of the tumor size, stage, age at diagnosis, and the histological appearance. According to one of the reports in literature out of 15 reported cases of teratoid Wilms’ tumor, chemotherapy was given in 9 cases and only 1 case showed cytoreductive response. Resistance to chemotherapy and radiotherapy is thought to be due to the presence of well-differentiated heterologous components. In the present case, as the tumor was limited to kidney and has been completely excised (Stage I), no post-operative chemotherapy was given after reviewing the literature. As there is still controversy regarding treatment, further, research should be carried out before concluding a tight treatment protocol in these cases.
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How to cite this article: Sinha A, Phukan JP, Bandyopadhyay G, Mukherjee S. Teratoid Wilms’ tumor in a child: A report of a rare case. Int J App Basic Med Res 2013;3:72-4.

Source of Support: Nil. Conflict of Interest: None declared.