

Adult teratoid Wilms' tumor- a rare entity with review of literature

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Abstract: Teratoid Wilms' tumor is a rare variant of Wilms' tumor (WT) which mostly occurs in paediatric age group. We are describing here a rare case of renal mass with teratoid histology in an adult female. The tumor was removed by enucleation. Grossly, the removed tumour was well-encapsulated homogeneous mass measuring 2 x 3 cm. On microscopy triphasic patterns of WT and prominent heterologous components were seen, including rhabdomyoblasts, bone, cartilage, and various types of mature epithelium. This is probably third reported case of a teratoid WT occurring in an adult kidney.

Keywords: Wilms' tumor, Nephron sparing surgery, teratoid histology

INTRODUCTION

Wilms' tumor is a common primary renal tumor, named after the German surgeon Dr. Carl Max Wilhelm Wilms, also commonly termed as nephroblastoma [1]. The term teratoid Wilms' tumor is used to describe an unusual variant of nephroblastoma in where predominantly heterologous tissues are found [2]. Classical triphasic combination of blastemal, stromal and epithelial cells are present in majority of lesions, occasionally other heterologous elements like squamous or mucinous epithelium, smooth muscle,

adipose tissue, cartilage, and osteoid and neurogenic tissue are also found. Fewer than 30 cases of teratoid Wilms' tumor have been reported until date [3]. Most of them were described in paediatric patients. Here we are describing a rare adult case of Wilms' tumor with teratoid histology.

CASE REPORT

A 22-year-old female came with an incidentally detected 2cm x 3cm right sided renal mass on Ultrasonography [fig1].



Fig-1: Ultrasonography showing right renal mass

Computer tomography scan revealed 2cm x 3cm hypovascular right sided hilar mass [fig2]. Pre-operative routine investigations were normal. With a provisional diagnosis of small renal mass of right

kidney, the tumor was extirpated by enucleation and the specimen was sent for the histopathological examination.

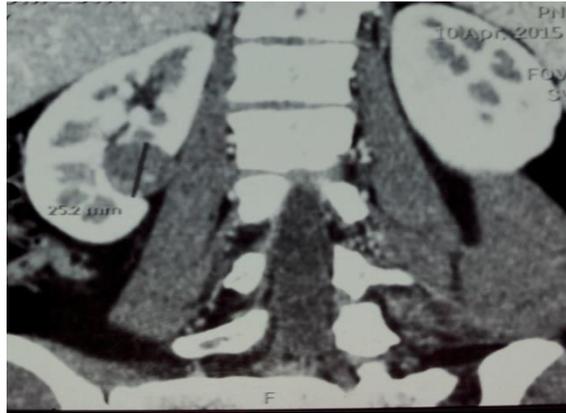


Fig-2: Computer tomography scan showing hypovascular right hilar mass

On gross examination, the tumor measured 2cm x 3cm. It was well-circumscribed and encapsulated. Cut sections presented homogenous gray-

white tumor tissue, soft and granular in the texture [fig3].



Fig-3: Cut sections of the right renal mass

Microscopically multiple sections examined from different areas showed classic triphasic combination of blastemal, stromal, epithelial

component along with cartilage, bone and rhabdoid tissue [fig4].

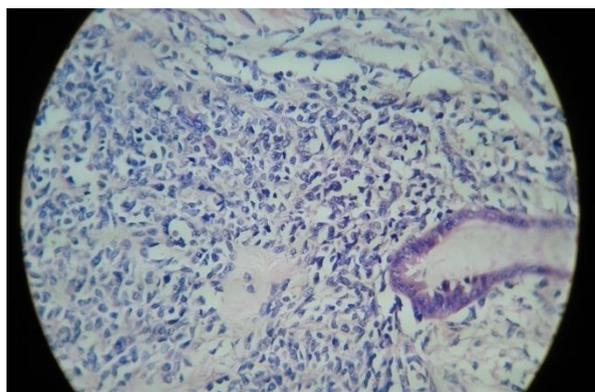


Fig- 4: Microscopic examination showing triphasic pattern of blastemal, stromal, epithelial component

Considering the above histopathological features, immunohistochemistry of WT1 gene done and

which was positive and a diagnosis of teratoid Wilms' tumor was arrived [fig5].

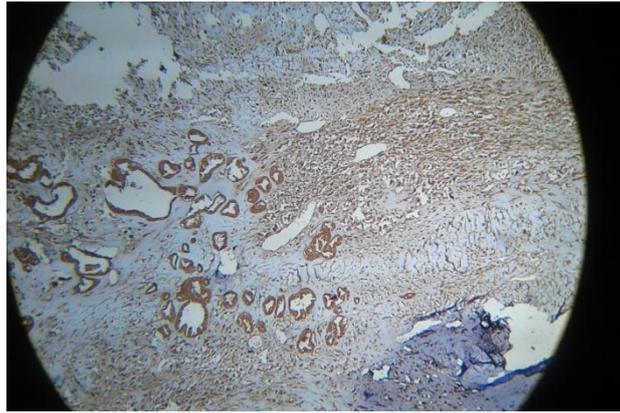


Fig-5: Positive immunohistochemistry of WT1 gene

No post-operative chemotherapy was advised. Patient was doing well in one year post-surgery follow up.

DISCUSSION

Wilms' tumor accounts for 0.5% of all renal neoplasms in adults [4]. Fifty percent of these tumors occur before 3 years of age and more than 90% of these are seen before the age of 6. Kidney is the most affected organ and both sides are equally affected. The term "teratoid Wilms' tumor" was first used by Variend *et al.*; in 1984 [5]. In 1988, Fernandes *et al.* proposed the criteria of presence of more than 50% heterologous component for the diagnosis of teratoid Wilms' tumor [2]. Teratoid Wilms' tumor is a rare histological variant of classical Wilms' tumor, with a predominance of teratoid elements [2, 6]. The pathogenesis of this rare condition is still unknown; probably the heterologous tissue components arise from primitive metanephric blastema [7]. Until date, fewer than 30 cases of teratoid Wilms' tumor have been reported [3]. Most of the patients were diagnosed between 2 to 4 years of age and 6 bilateral cases were observed [6]. Only two cases occurring in an adult has been reported until now [8].

Teratoid Wilms' tumor is usually not aggressive with a favourable outcome [2, 9]. Because of its rarity and presence of varying tumor components, uniform treatment regimen of this tumor has not yet been defined. Teratoid Wilms' tumor is relatively resistant to chemo and radiotherapy and surgery is the treatment of choice [2, 3, 9]. Resistance to chemotherapy and radiotherapy is thought to be due to the presence of the heterologous components [6].

CONCLUSION

In our case, the tumor was limited to kidney and has been completely excised, no post-operative chemotherapy was advised. In summary, we have reported a probably the third case of adult teratoid WT in 22-year-old female. Patient is in follow up since last 1 year without any recurrence.

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