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Available online at: http://www.iajps.com *A Case Study*

A RARE CASE OF A BLASTEMAL-TYPE WILMS' TUMOR IN A 4-YEAR-OLD BOY

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Abstract:

Wilm's tumor is a common malignancy amongst children, it has multiple histologic subtypes that mostly have a high cure rate when treated by resection and chemotherapy, however, a particular subtype which is called the "Blastemal-type" Wilms' tumor occurs less frequently in children and requires more intensified regimens of treatment. Herby, we present a case of a young boy who was histologically diagnosed with Blastemal-type Wilms' tumor.

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INTRODUCTION:

Wilms' tumor, also known as nephroblastoma, is one of the most prevalent tumors in pediatric age groups accounting for almost 6% in pediatric associated malignancies. Approximately 75% occur in ages below 5 years with a median of 3 years[1].

Wilms' tumor prevalence also varies with ethnicity groups being more common in the African Americans and less common in the Asian population[2]. Moreover, gender variance was noted as it is slightly more common in female compared to males. Furthermore, it is predominantly a sporadic disease with only 1-2% of patients with a positive family history, 10% of patient's have associated syndromes such as WAGR, Denys-Drash, and Beckwith-Wiedemann syndrome. With recent advances in screening, recognizing and management of the disease, "survival rates have increased to more than 90%, with re-occurrence rate of 15% in the population of a favorable histological pattern"[3].

Most important prognostic factors are the tumors' stage, molecular and genetic markers and histological subtype [4]; hence we would like to present this case that came along as a rare histological presentation.

Case:

A 4-year-old male presented to the clinic with his mother who has noticed a rapidly growing mass in the left side of his abdomen for the past year. She has also noticed a pinkish tinge to the color of his urine and noted that he complains of some pain during urination. On physical examination, he appeared generally fit and well, however, when the paediatric physician palpated the boys' abdomen, a large left-sided mass with smooth, regular margins that did not cross the midline was felt. The rest of the physical examination was unremarkable. Ultrasound and contrast-enhanced computed tomography (CT) scans of the abdomen were ordered. Ultrasound revealed a 15 x 13 cm well-defined mass of heterogeneous echogenicity arising from the left kidney. Heterogeneity is due to haemorrhage and necrosis. On CT scan, a large mass with low-density areas signifying tumor necrosis was seen. There was no evidence of nodal or hepatic metastasis. The right kidney was unremarkable.

The young boy was scheduled for an elective procedure and the left kidney was resected and sent to the laboratory. On gross examination, a lobulated tan mass with surrounding pseudocapsule is visualized arising from the left kidney. It appears as a varigated mass with prominent foci of necrosis and hemorrhage. Tissue from the tumor was submitted for histological examination. Usually, Wilms' tumor will demonstrate a mixed pattern of blastemal, stromal, and epithelial elements (Fig.1), however, it was different in our case.

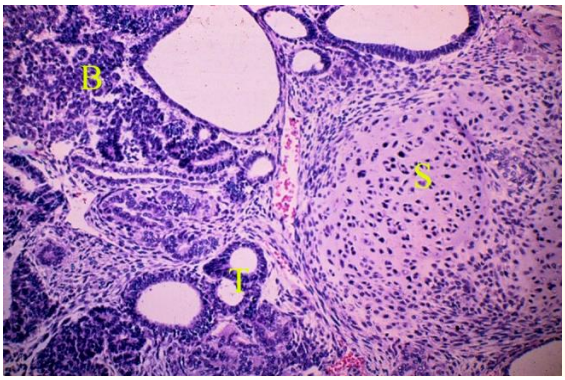


Figure. 1: Microscopic image of the classic Wilms' Tumor; the usual appearance of the triphasic tumor composed of blastema (B), epithelial elements (tubules) (T), and stroma (S). H&E, Intermediate power.

Microscopic examination revealed the following: the tumor is comprised entirely of blastemal component (Fig.2). No epithelial or stromal differentiation seen. No anaplasia seen.

Tumor was unifocal with a size of 155 mm in maximum diameter, the integrity of Gerota's fascia was intact (Fig.3). The renal sinus was involved with a more than minimal extension into renal sinus soft tissue. No renal vein involvement, extension beyond renal capsule, direct extension into adjacent organs or nephrogenic rests were identified. All margins and all regional lymph nodes were negative for tumor.

Pathologic Stage: Children's Oncology Group Staging System for Pediatric Renal Tumors Other Than Renal Cell Carcinoma: Local Stage II: Tumor more than minimally involves the renal sinus soft tissue. Immunohistochemistry results: WT-1, Pan-CK: Positive Vimentin, Synaptophysin, CD-99: Negative (Fig.4-5).

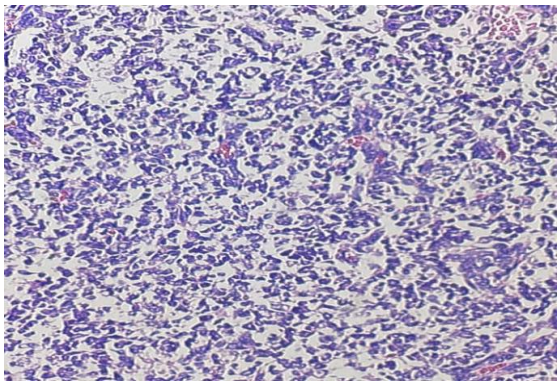


Figure. 2: Microscopic image of Blastemal-Type Wilms' Tumor; Tumor composed of entirely blastemal cells. H&E, Intermediate power.

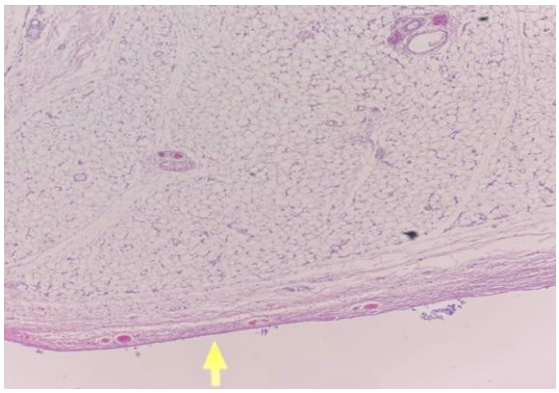
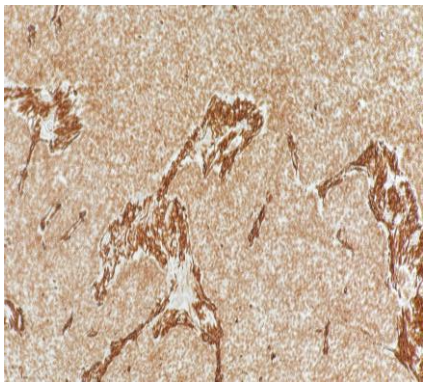


Figure. 3: Microscopic image of the intact Gerotas' Fascia. H&E, Low power.



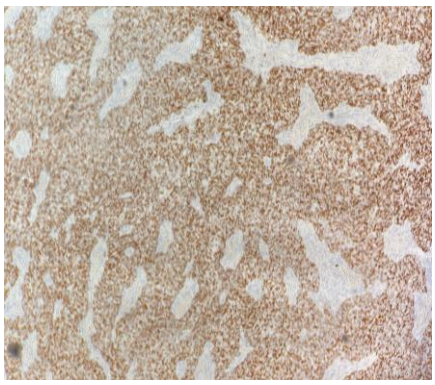


Figure. 4&5: Microscopic images of the Positive Immunohistochemical Stains; WT and Pan-Cytokeratins respectively. H&E, Intermediate power.

DISCUSSION:

Wilms' Tumor is the most common renal malignancy in children, and its treatment is one of the great success stories in pediatric oncology with cure rates approaching around 90%. This improved outcome has been the result of development of biological and clinical prognostic factors, which have guided for risk directed therapy[5]. Classical histological features of WT include a triphasic pattern of epithelial, stromal and blastemal components. The proportions of these

components and their lines and degree of differentiation vary significantly, resulting in countless tumour appearances. Biphasic and monophasic variants are not uncommon[6]. A staging system is a standard way for the cancer care team to sum up the extent of the tumor. In the United States, the Children's Oncology Group staging system is used most often to describe the extent of spread of Wilms tumors. This system divides Wilms tumors into 5 stages using Roman numerals I through V. Stage I: The tumor is contained within one kidney and was removed completely by surgical resection, stage II: The tumor has grown beyond the kidney, either into nearby fatty tissue or into blood vessels in or near the kidney, but it was removed completely by surgery without any apparent cancer left behind, Stage III:

This stage refers to Wilms tumors that most likely have not have been removed completely, Stage IV: The cancer has spread through the blood to organs away from the kidneys such as the lungs, liver, brain, or bones, or to lymph nodes far away from the kidneys, Stage V:

Tumors are found in both kidneys at the time of diagnosis[7]. The other main factor in determining the prognosis and treatment for a Wilms tumor is the tumor's histology, which is based on how the tumor cells look under a microscope. The histology can be either favourable or anaplastic. Microscopic structure of nephroblastoma or more commonly known as Wilms' tumor is associated with prognosis, and it us allows to distinguish 3 groups of tumors according to their malignancy: 1) low-risk group, which includes cystic partially differentiated and completely necrotic nephroblastoma; 2) intermediate-risk group – epithelial, stromal, mixed, regressive types and nephroblastoma with focal anaplasia; and 3) high-risk group – blastemal type, nephroblastoma with diffuse anaplasia[8].

Therapeutic options at presentation, based on stage, consist primarily of surgical resection followed by adjuvant chemotherapy or radiation therapy in patients with more advanced disease[9]. The patients with blastemal predominant tumors demonstrated a significantly worse prognosis compared with those of other subtypes. Hence, the treatment strategy of blastemal predominant category should be distinguished from the other favorable subtypes. The use of intensive chemotherapy is indicated in the case of blastemal-type Wilms' tumor even though it has a high failure and recurrence rate[10].

CONCLUSION:

Significant improvement has been made in the treatment of children with Wilms' tumor with new protocols that are designed to maintain a high rate of cure for these patients while minimizing toxicity, based on refinement of the risk stratification system. It is imperative that surgeons understand the directives of these new protocols and how the conduct of an operation can influence the outcome for these patients.

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