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WILMS TUMOR

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Abstract*:*

If the early cells of the kidney do not develop into glomeruli or nephrons, clusters of the immature cells may form in the kidneys when the baby is born. Usually, these cells mature by the time a child is 3 or 4 years old, but some may grow out of control, forming a mass of immature cells. This mass is called a Wilms tumor. It can also be called a nephroblastoma.

A Wilms tumor is always cancerous and is the most common type of kidney cancer diagnosed in children. A cancerous tumor is malignant, meaning it can grow and spread to other parts of the body. However, a Wilms tumor is very different from adult kidney cancer.

Wilms tumor usually occurs in only 1 kidney, called unilateral. However, it can develop in both kidneys, called bilateral. Rarely, a Wilms tumor develops in 1 kidney first and then the other.

A Wilms tumor is often found only after it has grown to a size of about 8 ounces, which is about 4 times the weight of a healthy 3-year-old child's kidney. About 1 out of 4 children with Wilms tumor have evidence that the tumor has spread either to the lung or liver when it is first diagnosed.

In this report will disscuss.

Introduction:

Wilms tumor (nephroblastoma), an embryonal malignancy of the kidney, is the most common renal tumor of childhood. It usually presents as an abdominal lum in an otherwise apparently healthy child. Abdominal pain, fever, anemia, hematuria, and hypertension are noted in 25%-30% of affected children. Approximately 5%-10% of individuals with Wilms tumor have bilateral or multicentric tumors. The prevalence of bilateral involvement is greater in individuals with a predisposition to Wilms tumor than in those without a genetic predisposition. A definitive diagnosis of Wilms tumor can be made only on histologic assessment of the tumor.Nephrogenic rests, benign foci of embryonal kidney cells that persist abnormally into postnatal life, are considered to be Wilms tumor precursors. Pathogenic variants may predispose to nephrogenic rests. Additional pathogenic variants transform nephrogenic rests into a Wilms tumor.

Discussion:

-Wilms tumors are the most common cancers in children that begins in the kidneys. About 9 of 10 kidney cancers in children are Wilms tumors. Most Wilms tumors are unilateral, which means they affect only one kidney. Most often there is only one tumor, but 5% to 10% of children with Wilms tumors have more than one tumor in the same kidney. About 5% of children with Wilms tumors have bilateral disease (tumors in both kidneys). Wilms tumors often become quite large

before they are noticed. The average newly found Wilms tumor is many times larger than the kidney in which it started. Most Wilms tumors are found before they have spread widly (metastasized) to other organs. Even when a doctor thinks a child might have a cancer such as Wilms tumor based on a physical exam or imaging tests, they can't be sure until a small piece of the tumor is checked % diagnosed by the lab. In 10%-15% of personnel with Wilms tumor, the cause is considered to be a germline related pathogenic variant or an epigenetic mutation occurring early during embryogenesis. These may or may not be related with a known congenital malformation syndrome or hereditary cancer syndrome. Approximately 1%-2% of these individuals with Wilms tumor have at least one relative also diagnosed with Wilms tumor (familial Wilms tumor); however, while germline variants that are likely pathogenic have been identified in some families, they are still unknown for the major classes of individuals. Among eight individuals reported with mosaic variegated aneuploidy and germline monoallelic BUB1B variants, seven (87.5%) had Wilms tumor. However, only about ten individuals with mosaic variegated aneuploidy and Wilms tumor have ever been reported.

Wilms tumor associated genetic abnormalities WT1 gene, dominant oncogene (at chromosome 11p13), WT2 gene (at chromosome 11p15).

The gene WT1 associated other syndrome, wagr syndrome and denys-drash syndrome.

WAGR syndrome (wilms tumor , aniridia) syndrome,, aniridia. Found 1.1% of wilms tumor patients, is caused by abnormality of the pax6 gene located adjacent to wt1 gene.

Wilms tumor develops in 40% to 70% of aniridia patients with deletions of wt1 gene.

Denys- drash syndrome (DDS) caused by point mutations in zinc finger DNA binding region of wt1 gene ,, 90% of DDS patients harbor germline point mutations is only one wt1 allele gene.

CONCLUSION*:*

Wilms tumor is the most common primary renal tumor of childhood and the fourth most common pediatric malignancy in the United States. It arises in approximately 10 children per million under age 15 years and is usually diagnosed between ages 2 and 5 years. Approximately 5% to 10% of Wilms tumors involve both kidneys, either simultaneously *(synchronous)* or one after the other *(metachronous)*. Bilateral Wilms tumors have a median age of onset approximately 10 months earlier than tumors restricted to one kidney.

References:

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