CASE REPORT

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Sudden Death due to Undiagnosed Wilms' Tumor in an Adult*

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ABSTRACT: Sudden unexpected deaths due to natural causes constitute a large number of cases encountered by the forensic pathologist. In a majority of such cases, heart disease is responsible for sudden death. Rare disease entities resulting in sudden death are occasionally encountered and may not fit the classic epidemiological profile. We present a case of sudden death due to a previously undiagnosed Wilms' tumor (WT) in an adult. The pathology of WT is discussed, as is the topic of sudden death due to previously unrecognized malignancy.

KEYWORDS: forensic sciences, sudden death, malignancy, Wilms' tumor

Case History

A 29-year-old white male was found semi-conscious on the bathroom floor of his apartment by his roommate. The roommate tried to convince him to go to the hospital, but he refused. Suddenly, he became unresponsive and apneic. Emergency medical services was called, cardiopulmonary resuscitation was initiated, and he was transported to the hospital. Resuscitation was unsuccessful, and he was referred to the medical examiners office for autopsy.

In the two months prior to his death, the decedent had been to a physician several times complaining of intermittent right flank pain. Physical examination was negative. Urinalysis revealed micro-hematuria. The decedent had been treated with doxycyline, but the hematuria had persisted.

At autopsy, a 5.0 × 6.0 × 8.0 cm mass was found in the right kidney. The mass was tan to pink, partially necrotic, and appeared to originate in the upper pole. The mass infiltrated into the renal pelvis and lower pole (Fig. 1). Multiple tumorous nodules were identified in both lungs. The remainder of the gross examination was unremarkable.

Microscopic examination of the renal mass revealed small hyperchromatic cells in a sarcomatoid pattern with foci of tubules, consistent with Wilms' tumor (nephroblastoma) (Fig. 2). The nodules in the lungs represented metastases.

Toxicology was negative, as were serological studies for HIV, Hepatitis B and C. The cause of death was Wilms' tumor of the kidney with pulmonary metastasis. The manner of death was natural.

Discussion

Wilms' tumor (WT), or nephroblastoma, is a tumor primarily of young children, with 90% of the cases occurring before the age of six years (1). Cases of WT in adolescents and adults have been documented, but they are rare. WT may be associated with multiple congenital syndromes such as Wilms' tumor-aniridia-genital anomaly-retardation (WAGR) syndrome, Beckwith-Wiedemann syndrome and Denys-Drash syndrome. Specific genetic abnormalities have been identified in WT: the WT1 gene (chr 11p13) and WT2 gene (chr 11p15.5). The WT2 gene is found in patients with Beckwith-Wiedemann syndrome (1).

The classic clinical presentation in children is an abdominal mass discovered by the primary caregiver; hematuria, pain, and hypertension are less common signs and symptoms in this age group.

Approximately 250 cases of WT have been reported in adults. In their study of WT in adults, Kilton, Matthews and Cohen reduced the number of reportable cases from 192 to 35 due to inadequate documentation, such as lack of or unconvincing photomicrographs or incomplete histologic descriptions (2).

In adults, WT usually presents with flank pain, a palpable mass and hematuria (3). The pathologic diagnosis can be difficult because the blastemal component can be confused with many adult undifferentiated tumors. Extra-renal cases have been reported in adults (4,5). The survival rate is lower in adults than in children (6). The lung is the most common site of metastasis.

Grossly, tumors are usually solitary, well-circumscribed, soft, pale gray to tan and may contain areas of cystic change, necrosis or hemorrhage.

Microscopically, WT consists of undifferentiated blastema, mesenchymal (stromal) tissue and epithelial tissue. The epithelial component may form tubules or glomeruloid structures. Tumors may be biphasic (consisting of only two components) or monophasic (one component). The mesenchymal component may differentiate; the most common heterologous cell type is skeletal muscle (1).

Sudden death due to previously unrecognized malignancy is not common, but it is not considered rare. As would be expected, a majority of such cases occur in older adults, the population at most

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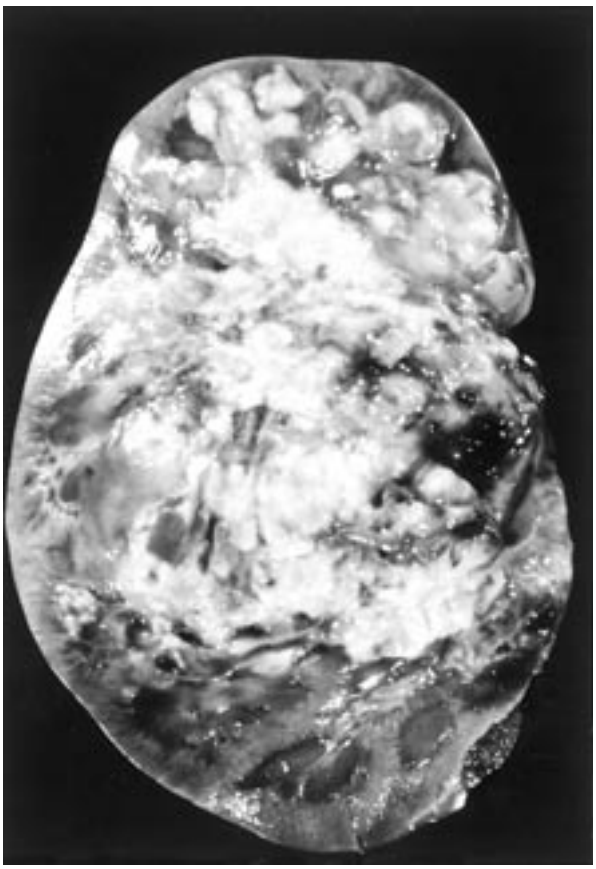

FIG. 1—*Gross photo of bisected right kidney.*

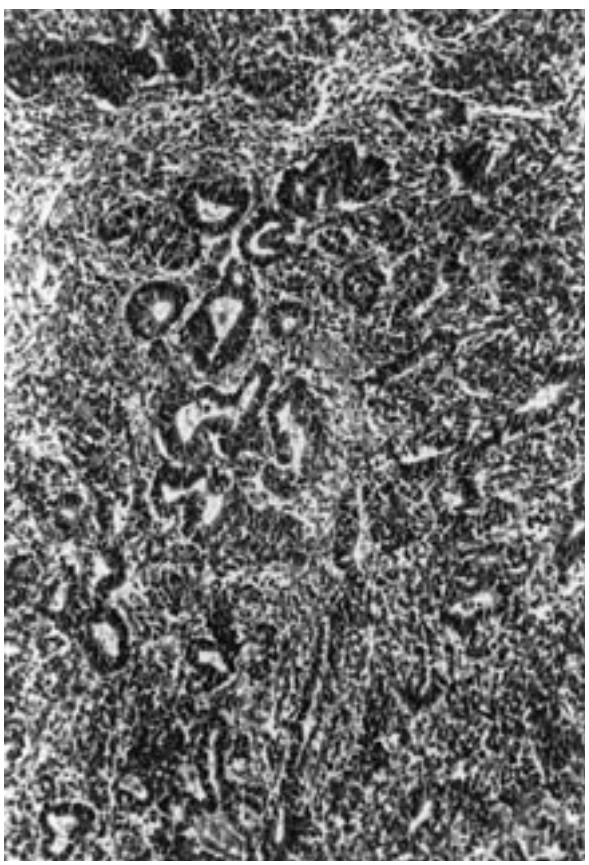

FIG. 2—*Low-power view of Wilms' tumor (H and E).*

TABLE 1—*Undiagnosed malignancies causing sudden death in young adults.*

Tumor Type	Number of Patients
Bronchogenic carcinoma	2
Pancreatic carcinoma	2
Acute leukemia	2
Adenocarcinoma of the urinary bladder	1
Astrocytoma	1
Carcinoma of the ventricle with widespread metastasis	1
Gastric carcinoma	1
Papillary carcinoma of the thyroid gland	1
TOTAL	11
(Results tabulated from references 7,8,9)	

risk for malignancy. Sudden death from undiagnosed malignancy is much less common in younger adults. We reviewed three series of sudden death in young adults (less than 50 years) and found that only 11 out of 430 individuals (2.6%) died as a result of undiagnosed malignancies (7–9). None of these malignancies were of renal origin. The most common primary sites were the pancreas, lung and bone marrow (Table 1). In studies that include older adults, carcinomas of the lung, pancreas and stomach are the most common previously unrecognized malignancies reported as causing sudden death (10,11).

Summary

Cases of Wilms' tumor in adults are rare, as are cases of sudden death due to primary renal tumors. The present case represents the first reported case of sudden death of an adult due to a previously undiagnosed Wilms' tumor.

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