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PRIMARY EXTRARENAL WILMS' TUMOR IN THE RETROPERITONEUM

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- **ABSTRACT**

Extrarenal Wilms' tumor is extremely rare. Approximately 54 cases have been reported in the world literature through 1993.Only eight extrarenal Wilms' tumors were enrolled in the National Wilms' Tumor Study (NWTS) between 1980 and 1986. These cases constituted a fraction of 1% of all Wilms' tumors reported to the N.W.T.S during this period. The authors report a case of retroperitoneal Wilms' tumor (Stage III) presenting as a lower abdominal mass in a previously healthy 6-year-old girl who was operated and staged according to the current N.W.T.S III criteria.

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Key Words • Wilms' tumor, extrarenal • nephroblastoma • retroperitoneal space

Introduction

Wilms' tumor or nephroblastoma is one of the most common malignant tumors of childhood.[1] It is most commonly a unilateral disease, but in 5% to 10% of the cases both kidneys are affected.[2] Extrarenal tumors are extremely rare and have been the subject of isolated case reports.[3] When the tumor arises in the retroperitoneal space, uncertainty may arise as to whether they are truly extrarenal in origin or a secondary extrarenal metastases.[3] The diagnosis of extrarenal Wilms' tumor can be justified by appropriate radiologic, surgical and pathologic criteria.

Herein, a case of an extrarenal Wilms' tumor arising in the retroperitoneum in the lumbosacral area, is presented and the literature reviewed. Probable tumor origins are also discussed.

Case Report

A 6-year-old girl was admitted to the pediatric surgery ward with the chief complaint of fullness in the lower abdomen. She was apparently well until two months prior to referral, when she developed gradual abdominal pain, poor appetite and severe weight loss. Her family history was negative.

Examination, revealed an anemic, malnourished patient with a palpable fixed smooth mass of 10? 10 cm in hypogastrium extending from the umbilicus to the pelvic region.

Laboratory investigations were as follows: hemoglobin 8.5 g/100 ml, hematocrit 25%, white blood cell count 11000/mm^3, and erythrocyte sedimentation rate, 25 after first hour. Urinalysis, serum BUN and creatinine were normal.

Chest X-ray was normal and plain X-ray of the abdomen revealed displacement of the bowel gas to the right upper side. Intravenous pyelogram showed a left-sided hydronephrosis, lateral displacement at the left ureter and fullness of the right side. CT scan showed a mass in the lower abdomen with foci of hypodensity in the center suggestive of necrosis. No abnormality was detected inside the kidneys (Figs.1,2).

Exploration revealed a mass located in the retroperitoneum starting from lumbosacral area to pelvis. Small bowel mesentery, left ureter, abdominal aorta, and left iliac vein were involved. Thus, the tumor was stage III. Because of invasion of these vital organs, only debulking of the mass could be performed, with gross evidence of residual tumor tissue remaining in the region (Figs. 3). Despite extensive chemotherapy and local radiotherapy, the patient passed away 13 months after the initial diagnosis.

Discussion

Wilms' tumor or nephroblastoma is one of the most common malignant tumors of childhood.[1] It is most commonly a unilateral disease. But in 5% to 10% of the patients both kidneys are affected.[3] Extrarenal cases are extremely rare and have been the subject of isolated case reports.[4] The diagnosis of extrarenal Wilms' tumor can be justified by appropriate radiologic, surgical and pathologic criteria. The diagnosis of extrarenal Wilms' tumor is made only after a primary renal tumor has been ruled out.[4]

The histology of extrarenal Wilms' tumor is similar to that of intrarenal Wilms' tumor, nevertheless the exact embryological origin of the latter is not clear. The location of extrarenal Wilms' tumor arising from a position as cranial as the chest wall to as caudal as the scrotum raises the possibility that the lesion might arise from a more primitive mesonephric or pronephric origin.[2,5]

A Wilms' tumor that is adjacent to the gonads may have a mesonephric origin. The pronephros, the first and embryologically most primitive excretory organ, arises in a cranial position adjacent to somites 9 to 12, which are lower thoracic in position.[4]

Malignant degeneration of an aberrantly located cells derived from this tissue may account for a chest wall tumor. Coppes et al[3] have performed an extensive literature review of extrarenal Wilms' tumor among the pediatric and adult populations, which was reported through 1990. There were 34 cases, thirteen of which had tumors in the inguinal canal, scrotum or round ligament.

In the females, in that series, the inguinal region or round ligament was the most common site. However, the most common site among the males, as in this patient, was retroperitoneum.[6]

Clinical course of the extrarenal Wilms' tumor parallels those of intrarenal ones. It should be, however, staged and treated according to current N.W.T.S. protocol. Higher staging in these cases (usually stage III) may be explained on the basis of rare occurrence of the tumor in unusual locations, causing delay in diagnosis and necessary work-ups. Fine needle aspiration with preoperative chemotherapy could have been another option before exploration. Thus clinical suspicion is the main determinant factor if diagnosis is not to be missed.

Pathologic Finding

Sections form the excisional mass showed necrosis and was composed of blastema and mesenchymal tissue.

The blastematous areas are extremely cellular and composed of small round-to-oval primitive cells, the cytoplasm being very scanty (Fig. 3).

The pattern of growth is nodular cord-like or showing epithelial embryonic tubular formation and immature glomeruli. The mesenchymal elements have spindle cell fibroblast-like configuration.

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