Case Reports

Extrarenal Wilms' Tumor

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Introduction

Extrarenal Wilms' tumor, which by definition excludes primary tumor in the kidney, is extremely rare. It occurs predominantly in children. Most of the cases that have been reported involved the retroperitoneum.

We are reporting a case a of retroperitoneal (extrarenal) Wilms' tumor extending into the left psoas muscle and through neural foramina into the spinal canal with meningeal metastasis in a neonate. To the best of our knowledge, such an extensive extrarenal Wilms' tumor at this age with intraspinal extension and meningeal metastasis has not been reported previously

Case Report

An 8-day-old full term baby boy presented with jaundice, developed on the 3rd day of life, and vomiting with hematemesis since the 5th day of life. No antenatal ultrasound examination had been performed. The baby was full term and had a normal vaginal delivery.

Clinical examination revealed a firm and non-tender mass in the right iliac fossa. X-ray of abdomen revealed a mass of soft tissue density, occupying lower abdomen and pelvis, displacing bowel loops. The ultrasound examination showed a large, solid, predominantly hyperechoic mass, measuring 85 mm in longest dimension, with internal vascularity suggestive of neoplastic lesion. Posteriorly the mass was immediately adjacent to the spine. There was no connection of the mass to the kidneys. Mild hydronephrosis of the right kidney was identified. As the lesion was suggestive of malignancy, CT of abdomen was done which showed a heterogeneously enhancing, solid, retroperitoneal lobulated mass about 100x80mm in dimensions (Figure 1). The mass was extending from the level of lower pole of both kidneys, displacing them and pushing the aorta and iliac vessels anterolaterally. In addition, at the level of L2/3, the mass was encroaching into the spinal canal through the left sided neural foramen expanding it without bony destruction. Hydronephrosis of the right kidney was also noted. The rest of the abdominal viscera were normal. There

was no evidence of lymphadenopathy on CT examination. Differential diagnosis included neuroblastoma and teratodermoid tumor.

The child underwent exploratory laparotomy. The surgeons extracted a retroperitoneal mass of 10x8cm from the lumbosacral region which was extending upward with the aorta and iliac vessels stretched over the tumor. The tumor was extending into the spinal canal at the L2/3 level. Tumor site, extent and appearance resembled that of neuroblastoma and the intraspinal component was not removed. The operation was successful and recovery was uneventful.

On gross examination, the specimen consisted of a large nodular tissue measuring 9x7.5x5 cm. The cut surface was solid, lobulated and grayish white in color. Histopathological examination showed features of Wilms' tumor, exhibiting both epithelial and mesenchymal differentiation. The features of anaplasia were not appreciated (Figure 2). Resected lymph nodes showed benign lymphoid hyperplasia.

A magnetic resonance imaging (MRI) was not carried out preoperatively. Follow up MRI examination showed a 3 cm mass in the left psoas muscle extending through a left neural foramina into the extradural space from L4 to S1 (Figures 3a, b). Pial enhancement over the conus medullaris of the spinal cord was also identified suggesting meningeal metastasis (Figure 4). L4/5 laminotomy and removal of extradural tumor was carried out. The patient received chemotherapy (Vincristine and Actimomycin D). Follow-up to date has been over 1 yr and 10 months and the patient is currently well.

Discussion

Extrarenal Wilms' tumor is extremely rare. According to our review of the literature 72 cases have been reported[1-40] (including the present case report as shown in Table). These tumors can occur in isolation or in association with other tumors usually teratomas.

The diagnostic criteria necessary to establish the diagnosis include absence of primary kidney tumor and supernumery kidney (radiologically and surgically).

The origin of Wilms' tumor is controversial. An extrarenal location supports more frequent occurrence of ectopic metanephric blastema.[1] But, the presence of tumor cephalad to a horseshoe kidney favors origin from primitive mesodermal tissue, probably mesonephric rests.[2] Presence of persistent mesonephric duct remnants in the walls of cervix, vagina and inguinal canal and occurrence of extrarenal Wilm's tumor in these locations supports the theory of mesonephros as the source of origin.[3] Another possible mechanism involves Connheim's cell rest theory in which cells with persistent embryonal potential undergo malignant transformation.[3] These tumors can also arise from other neoplasms, generally teratoma.[2]

A palpable mass is the most common presentation. Patients with uterine extrarenal Wilms' tumor may present with irregular menstrual bleeding. Two patients with thoracic lesions were discovered on chest radiograph for nonspecific pulmonary symptoms.[4-6]

Age of presentation is reported to be between 2 months to 10 years. The oldest recorded patient suffering from uterine extrarenal Wilms' tumor was a 77 years old female.[7] It is more common in females, 56.6% of reported cases occurred in females and 43.4% in males.[1-40]

Extrarenal Wilms' tumors are mostly solid large masses. Cystic cavities associated with a solid tumor can occur.[13,14] One unusual manifestation presented from a large cystic component mimicked ascites clinically.[4] Extrarenal Wilms' tumor has the potential for local recurrence[15-17] and distant metastasis. Metastasis have been reported in lungs[15], liver, pancreas[18] and brain.[19] Venous invasion of a right suprarenal tumor into the IVC and right atrium has also been reported.[2]

Six patients of extrarenal Wilms' tumor were associated with horseshoe kidney, i.e., 7.6% of patients having extrarenal Wilm's tumor had horseshoe kidneys in association.[20] This tumor has also been reported in patients with spinal dysraphysm.[19,21]

Regarding staging and management of extrarenal Wilms' tumor, the behavior of this tumor appears to parallel that of a similarly staged intrarenal Wilms' tumor. Hence, the staging and management protocols of intrarenal Wilm's tumor can be applied to an extrarenal location.[22] An adaptation of the National Wilms' Tumor Study (NWTS) protocol can be carried out.[3] Currently TNM classification is used for staging purpose.[22]

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