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Primary Extrarenal Wilm's Tumor Associated with Horse Shoe Kidney- A Rare Case Report.

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Abstract

Nephroblastoma (Wilms' tumor) is the most common renal malignancy in childhood, however Extrarenal Wilms' tumor is extremely rare, mostly reported in retroperitoneal location. Diagnosis is almost always postsurgical as it often misdiagnosed for other common retroperitoneal masses of that region. Association with horse shoe kidney raises an important issue regarding the histogenesis of these tumors

We report a case of 4 years old boy presented to emergency with a complaint of abdominal pain, fever and vomiting of one week duration post abdominal trauma. Imaging reveals a large 8.5 cm retroperitoneal mass, encasing the right lower pole of a horse shoe kidney, associated with haemeperitoneum and compressing bladder, right ureter and small bowel

The child was taken for emergency laparotomy which revealed a well- encapsulated right-sided retroperitoneal tumour separate from the right kidney, however rupture. Tumor was completely excised, diagnosis confirmed by histopathology. Adjuvant chemotherapy was given as per protocol. No metastatic/recurrent disease detected in 2 years follow up.

Introduction

Wilms tumor (WT), also known as nephroblastoma, is the most widely reported primary malignant renal tumor in childhood, with a prevalence of eight per million children [1].

Horseshoe kidney (HK) is among the numerous renal anomalies that have been associated with WT [2]. A child with HK has a twofold increased risk of developing an embryonic tumor of the kidney compared with the general population of children without HK [3].

Extrarenal Wilms' tumors are exceptionally rare and have mainly been the subject of isolated case reports. The estimated rate of occurrence of nephroblastoma outside the kidneys is almost 0.5 to 1% of all cases of Wilms' tumor. Extrarenal Wilms' tumor (ERWT) occurs mostly in childhood; however, it is also rarely reported in adults [4]

The association of ERWT with a horseshoe kidney has been reported previously, and almost 13% of the reported ERWTs were found to be associated with the horseshoe kidney [5]

Nephroblastoma may be observed outside the kidneys in two other situations: metastatic disease and nephroblastoma arising in a teratoma. Hereby we present a rare case of primary extrarenal Wilms tumor in the retroperitoneum associated with horseshoe kidney.

Case Report

A previously healthy, 4 years old boy was admitted in the paediatric surgery department, with the complaints of complaint of abdominal pain, fever and vomiting of one week duration post abdominal trauma. The child was normotensive with no clinical dysmorphism. His growth parameters were on the 50th centile.

On examination his abdomen was distended with an approximately 10 x 10 cm sized non mobile mass was palpable in lower abdomen. The hematological parameters and routine investigation reports were normal. Tumor markers: alpha fetoprotein was within normal limit, lactate dehydrogenase is slightly elevated.

Abdominal ultrasonography demonstrated a heterogenous mass with cystic and solid areas measuring 9.7 x 8.1 cm arising from midline pelvis and extending in to lower abdomen, located superior to bladder and anterior to aorta. It is displacing the bowel loops and abutting the anterior abdominal wall. Right kidney was separately visualized and show mild hydronephrosis. Left kidney was normal. (Figure 1)

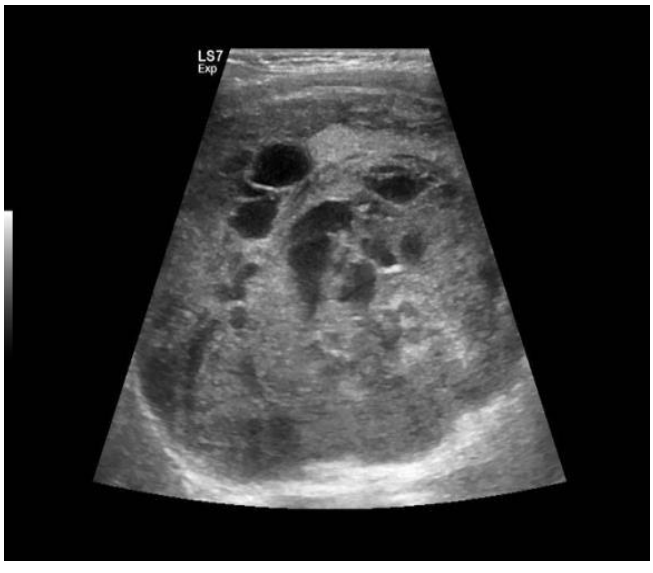

Figure 1: Ultrasound of the mass

CT scan of the abdomen showed an approximately 9.7x9.4x6.2 cm sized mixed density heterogeneously enhancing mass in the right retroperitoneal presacral location extending from L2 till second piece of sacrum, smoothly pushing the right ureter to anterolateral aspect with hydronephrosis of the right moiety and a horseshoe kidney. (Figure 2) There was no abdominal lymphadenopathy. The radiological differential diagnosis was teratoma or sarcoma. CT chest was within normal limits.

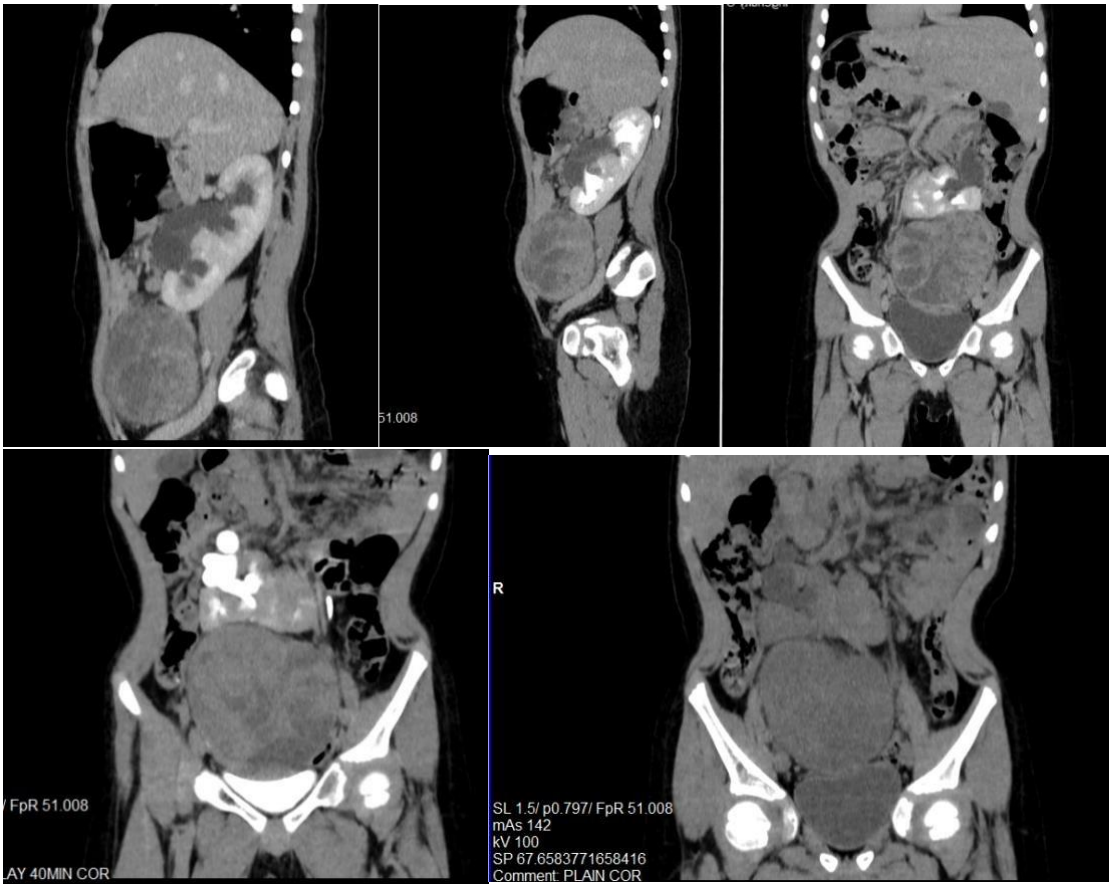

Figure 2: CT Scan: Heterogenous Mass with Horse Shoe Kidney

Emergency laparotomy with lower transverse abdominal incision performed, peritoneal cavity is opened, large mass seen, well capsulated retroperitoneal, ruptured just above the bladder with blood collection and small amount spillage of tumor content into peritoneal cavity and right iliac fossa. The bladder was compressed down and adherent to the tumor, the right ureter pushed antero-lateral. Suction of blood and fluid collection and tumor material was done, peritoneal wash performed, isolation of perforated area, then dissection and release tumor from the bladder and right ureter performed. The small bowel pushed upward and to the left side, the tumor abuts the lower pole of right kidney and was released from kidney without breaching the renal capsule or damaging the renal parenchyma, both right and left kidney seen joined together at lower pole. dissection posteriorly tumor seen just above major vessels and iliac vessels tumor crossing mid line and attached well to the spine L4,L5, S1,S2.

resection is done, tumor measure about 10x10cm. Peritoneal wash by saline done, no enlarged lymph nodes seen few lymph nodes taken from the mesentery of small bowel for biopsy. drain inserted wound closed in layer.

On gross examination a spherical encapsulated mass weighing 162 g and measuring 8.5 x 7.5 x 5.0 cm, external surface is smooth, the capsule is focally ruptured in some areas. Cut surface is hemorrhagic soft fleshy, whitish and greyish with cystic areas (Figure 3).

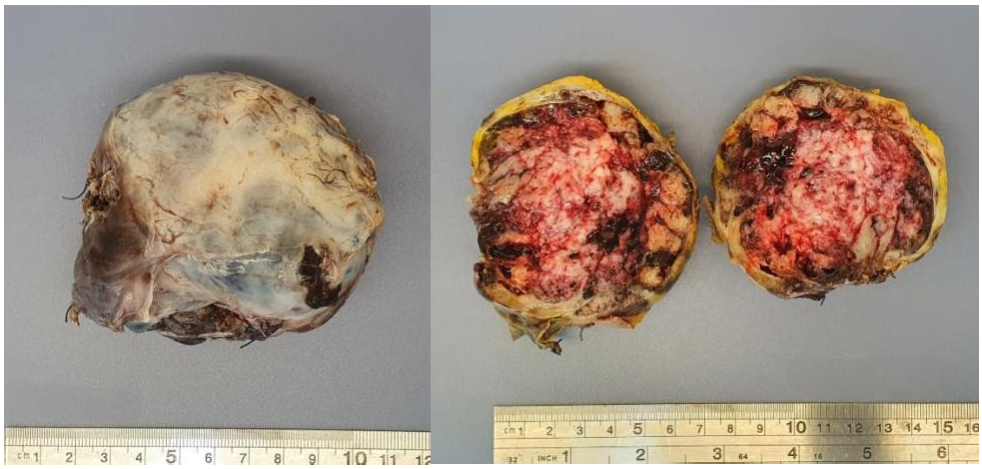

Figure 3: Gross pictures: Ruptured capsule with heterogenous cut surface

The mass extensively sampled to reveal a triphasic tumor composed of primitive blastemal component (40%). Epithelial component (20%), Stromal component (30%) range from hypo to hypercellular undifferentiated areas to well differentiated areas with focal skeletal muscle differentiation and minor component of heterologous elements (rhabdomyoblasts and adipose tissue). No features of anaplasia identified (Figure 4). Immunohistochemistry stains for WT1, P53 and Ki67 were performed with adequate contrls. WT1 is diffusely positive in the Plastema component, focally positive in the epithelial component and negative in the stromal component. Plastemal component shows high proliferation index with Ki67 (70%), while epithelial and stromal componentents are low (10%). P53 is completely negative (wild type). A diagnosis of Extrarenal nephroblastoma with favourable histology was given.

Tumor stage: III as per staging criteria for Wilms tumor in SIOP and COG due to tumor rupture prior to surgery. The examined lymph nodes were uninvolved by tumor.

Post operatively the CT abdomen showed no residual mass and intact horseshoe kidney (Figure 5).Post operative adjuvant chemotherapy was started to the child consisting of Dactinomycin, Doxorubicin and Vincristine in two courses along 28 weeks, combined with radiation on first and seventh weeks.

Child responded well to treatment with no recurrent or metastatic disease upon follow up for two years after diagnosis.

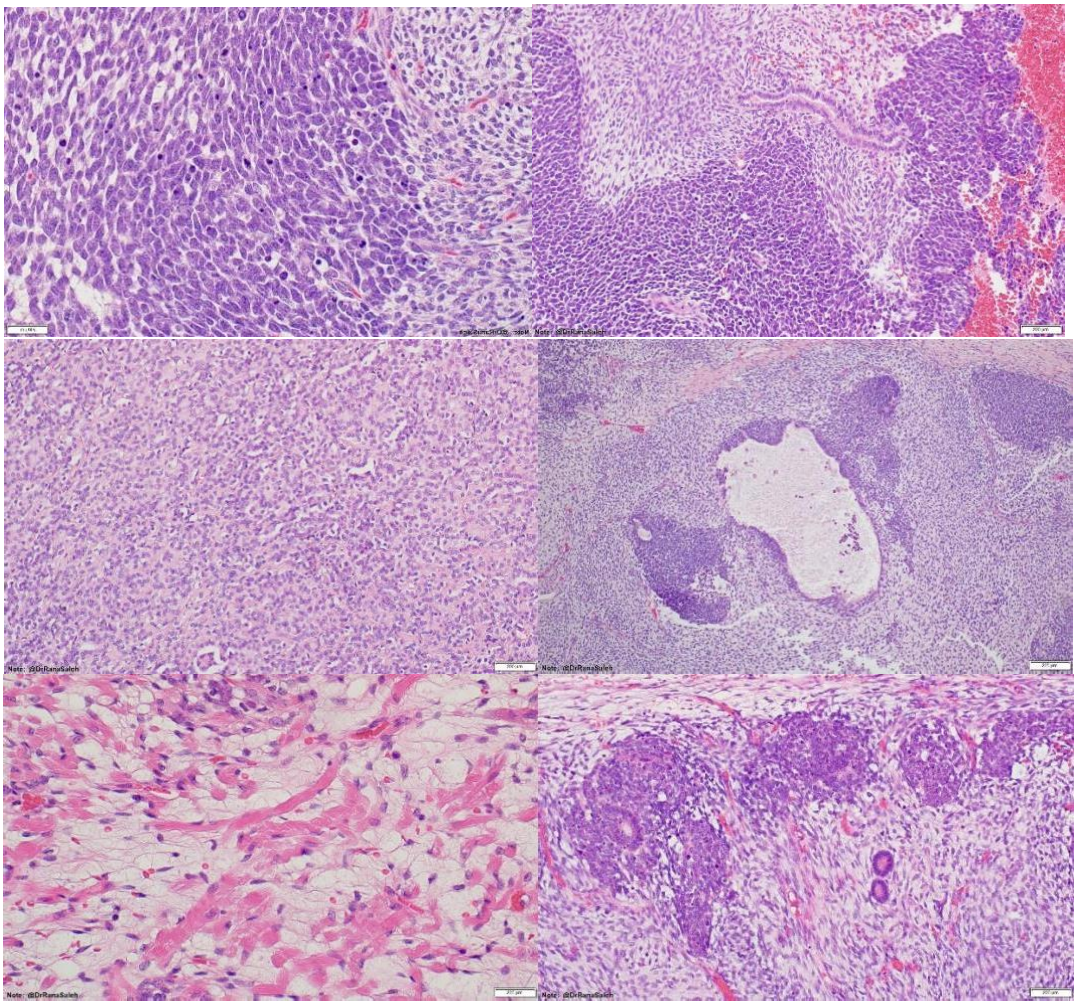

Figure 4: Microscopic pictures: Triphasic tumor with focal rhabdoid stromal differentiation

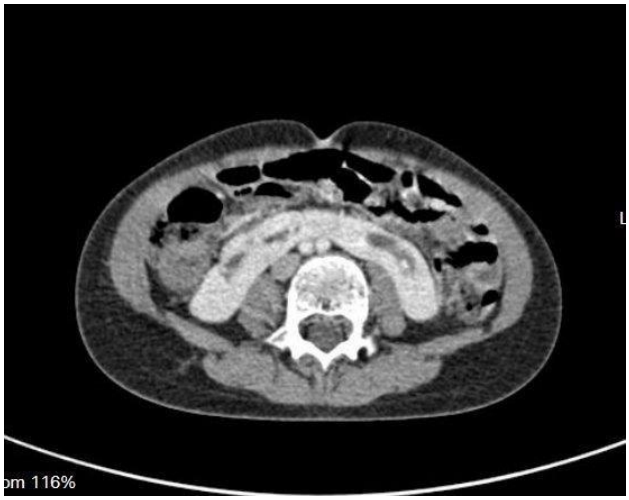

Figure 5: Postoperative CT abdomen shoes intact horseshoe kidney

Discussion

Wilms' tumour is a malignant neoplasm of primitive metanephric cells that retain their embryonic potential. The classical location for Wilms' tumour is the kidney. However, cases with typical morphologic features of Wilms' tumour have been recorded in extrarenal sites, including the retroperitoneum, sacrococcygeal region, testis, uterus, inguinal canal and mediastinum. [6]. The exact mechanism whereby a Wilm's tumor occurs in extrarenal tissues is unkown. However, cells of origin could be ectopic nephrogenic rests. The diagnostic criteria necessary to establish the diagnosis include: Absence of primary kidney tumor and supernumery kidney (radiologically and surgically) [7]. Our case is fulfilling these criteria in correlation with imaging studies and operation notes.

The imaging features of extrarenal wilm's tumor are nonspecific and always confused with other retroperitoneal tumors as mentioned in previous reports, however frequent association with genitourinary anomalies like horse shoe kidney should raise the possibility if done by expert. The diagnosis of ERWT proposed on imaging studies can be confirmed only upon pathological examination of the specimen.

nephroblastoma may be observed outside the kidneys in two other situations: metastatic disease and nephroblastoma arising in a teratoma. therefore, in the case of ERWT, it is mandatory to evaluate the kidneys for primary tumor preoperatively and search the whole specimen for any teratoid element postoperatively [5]. The classical Wilms' tumour has a triphasic histologic pattern, with blastemal, mesenchymal and epithelial components. The heterotopic mesodermal elements are commonly seen but usually involve a minor part of the neoplasm. According to Variend et al's criteria for diagnosing teratoid Wilms' tumour, the heterotopic elements should have dominant presence (> 50% of the total area) (6). In our case the mass was almost entirely submitted for microscopic examination and

The histology is favorable in the absence of anaplasia, heterologous elements are seen focally (approximately 3% of the total microscopic fields, upon extensive sampling) and did not qualify the criteria of teratoid Wilms.

Extrarenal Wilms is staged similarly to intrarenal counterpart, given stage II or higher since all of them are beyond the renal capsule. The staging system used in the literature while describing the case was International Society of Paediatric Oncology (SIOP) and Children's Oncology Group(COG), [13] our case is given stage III due to tumor rupture prior to surgery. The prognosis is also similar.

Surgical excision remains the key step in the treatment of ERWT, especially when performed radically [8] [13] Regional lymph node sampling is a part of the surgical principle as that for classic renal Wilms'

tumor. Careful inspection of solid organs such as kidneys or liver and also peritoneum for tumor implants is recommended in abdominal ERWTs. Chemotherapy as adjuvant therapy was reported to have been used in all studies [14]

Conclusion

ERWT is a rare malignant neoplasm with atypical presentations. This report suggests that ERWT must be kept in mind while confronting with an abdominal mass in a patient with horseshoe kidney ((11)The staging, response to chemotherapy and prognosis are similar to the renal Wilm's counterpart. Therefore, similar staging and treatment protocol can be followed.

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