

Case Report

Extra-Renal Wilms' Tumor in Left Lobe of Liver: A Case Report of the Rare Tumor with Review of Literature

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Abstract

Wilms tumor is the commonest childhood tumor. Extra renal Wilms' tumor is a nephroblastoma arising in locations other than kidneys. The tumor arises from misplaced metanephric blastema or is contemplated to arise from pluripotential cells. A metastasis, origin of tumor in kidneys and supernumerary kidneys have to be ruled out to assign a diagnosis of extrarenal Wilms' tumor. It has been commonly reported to occur in retroperitoneum but also in inguinal region, pelvis, testis and colon. The age of occurrence has been reported as Wilms' tumor in literature. It is mostly diagnosed post operatively. A preoperative biopsy combined with immunohistochemistry can help in

diagnosing tumors in cases where neoadjuvant chemotherapy is anticipated. The Wilms' tumors in extrarenal location largely have favourable histology and are treated on same guidelines as Wilms' tumor. Here we present a case of extra renal Wilms' tumor arising in left lobe of liver. Liver as location for an extra renal Wilms' tumor was not found in literature.

Keywords: Extra renal Wilms' tumor; Nephrogenic rests; WT1; Extra renal location

1. Introduction

Wilms' tumor is the commonest childhood tumor with incidence of one per 10000 children. The tumor usually

presents upto 15 years of age [1]. Incidence of extra renal Wilms' tumor is estimated to be 0.5-1% of all Wilms' tumors [2]. The presentation is progressive lump depending on location of tumor. The genesis of extra-renal Wilms' tumor is controversial. Two possibilities that need to be ruled out, are metastatic lump from a Wilms' tumor and teratoma. The tumor is believed to arise from mesonephric remnant or metanephric tissue. It may also arise from teratoma though this concept is not proven [3]. We present a case of a 5 year child with an extrarenal Wilms' tumor located in left lobe of liver as liver is not reported as site of occurrence for it.

2. Case Presentation

A 5-year-old child presented with abdominal distension with pain and laborious breathing. He had no bowel symptoms of vomiting or constipation. He appeared chronically malnourished. The pulse rate was 132/min and blood pressure of 104/60 mm of Hg. The abdominal examination revealed a lump occupying all quadrants of abdomen. Liver was not separately palpable and there was no ascites. He had normal external genitalia. Contrast enhanced CT examination of abdomen showed an abdomino-pelvic mass of 13.4 x18.4x20.2 cm, abutting left lobe of liver and stomach. Both kidneys were normal with no cyst, mass, hydronephrosis or lymph nodes. Chest CT scan showed a 3cm nodule on diaphragmatic pleura on left

side. Serum Alpha-fetoprotein levels were normal. The biopsy examination revealed a triphasic Wilms' tumor. Immunohistochemistry (IHC) was positive for WT1, CK, Bcl2, CD34 and CD99 in strong favour of Wilms' tumor. Patient was administered 8 cycles of neoadjuvant chemotherapy with weekly Vincristine 0.046mg/kg, five weekly, Adriamycin 2mg/kg and weeks 3 and 8 Actinomycin 45µg/kg. Patient was subsequently explored and found to have a tumor of 20x 14 x16 cm, densely adherent to or arising from left lobe of liver, free from diaphragm, parietal peritoneum and stomach. The diaphragmatic nodule indicated on CT scan was a 3 cm nodular extension from tumor abutting and indenting diaphragm. Both the kidneys were separately palpable and had no mass lesion, cyst or hydronephrosis. Tumor along with left lobe of liver was mobilized from diaphragm. Hilum of liver was dissected and left bile duct, left hepatic artery and left branch of portal vein was ligated and divided. Left lobe of liver along with tumor was excised along line of ischemia. Left hepatic vein was ligated and divided near diaphragm. The tumor was excised with the left lobe of liver. We found no attachments of tumor either to diaphragm, crus of diaphragm or the upper retroperitoneal area around bare area of stomach or aorta. Patient had uncomplicated recovery and was discharged on sixth post operative day. The tumor post operatively was reported on histopathology as Triphasic Wilms' tumor.



Figure 1: Clinical photograph

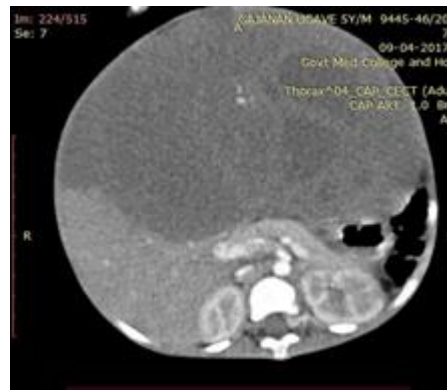


Figure 2: CECT abdomen- Left lobe liver mass with normal kidneys



Figure 3: Tumor in relation to left lobe liver



Figure 4: Postoperative Specimen

GOVT. LANCET HOSPITAL, SERANGARAI
DEPARTMENT OF ONCO-PATHOLOGY
HISTOPATHOLOGY REPORT

NAME: ELAJANAN UGAVE AGE: 5 YRS SEX: MALE
 REF. NO: 1690/22 Dept: 435-2 Ward: 2 Unit EC: Dr. ANR
 Specimen no: 1690/22 Request No: H-10555-22/22

SP-NO: 2080/22. Specimen labeled as: Tumor arising from left lobe of liver

Received single, globose mass of size 23x21x9 cm. Externally surface appears lobulated, multiple nodules seen. Weight of size 344.5x13.5 gm. On cutting open, soft cystic lesion identified with necrotic fluid oozing out. Normal liver identified of size 35x16.5 cm.

H-10555-22/22 and H-10556-22/22 - Sections stained show tumour tissue comprising of epithelial, fibrous and stromal elements. The epithelial element is composed of irregular, atypical tubules lined by elongated, stratified nuclei. The fibrous component is composed of small round blue cells with scanty cytoplasm, overlapping nuclei with finely dispersed chromatin. The ground component consists of spindle cells arranged in fascicles. Heterologous elements such as cartilage and bony elements seen. The absence of focal or diffuse anaplasia. The tumour is focally infiltrating into the liver.

H-10555-00/22 -Normal liver: Unremarkable.

IMPRESSION:
 Specimen labeled as: Tumor arising from left lobe of liver
 = TRIPHASIC WILMS' TUMOUR.
 *HETEROLOGOUS SQUAMOUS CELLS, CARTILAGE AND BONY ELEMENTS SEEN.
 * THE TUMOR IS FOCALLY INFILTRATING INTO THE LIVER.
 *NO FOCAL/DIFFUSE ANAPLASIA.
 Advice: IHC studies if indicated.
 kindly Issue blocks for IHC

PATHOLOGIST

Figure 5: Postoperative Histopathology

Name: **MR. ELAJANAN UGAVE** Clinic Details: **Lancet Hospital, Serangarai**
 Age: **5 Yrs** Sex: **Male** Department: **Pathology, Microbiology**
 Ref No: **13498/22** Acc. No: **000000017901**
 Regd. No: **0900000002** Build No: **000000000000**
 Cell No: **9199000000000000** Ward No: **0000000000000000**
 Ward. No: **0000000000000000** Hospital No: **0000000000000000**

IHC Final Diagnosis Panel

LAB. NO.: **01690/22**

CLINICAL DETAILS
 Clinical history: Right sided (supra of Abdominal mass). Suggestive of Triphasic Wilms' Tumor.
 CRIT T abdomen. A large heterogeneously enhancing soft tissue density abdominal/mesenteric mass lesion measuring 13.4 x 18.4 x 20.2 cm (AP x TR x CC) is noted in posterior cavity.

MSX - Abdominal mass

SPECIMEN DETAILS
 Received two paraffin blocks labeled as H-10555/22 & H-10556/22 for IHC.
 IHC performed on H-10555/22. Remaining unused as 0.

MORPHOLOGY
 Sections stained show tissue comprising of epithelial, fibrous and stromal elements. The epithelial cells is composed of the atypical tubules lined by elongated stratified nuclei. The fibrous component is composed of small round blue cells with scanty cytoplasm. Overlapping with finely dispersed chromatin. The ground component of spindle cells arranged in fascicles.

HC MARKERS	RESULTS
CK	Positive in tubules
AE1/AE3	Positive in tubules
CD45	Positive in stroma
CD45L	Negative in tumor cells
CD56	Positive
EMA	Negative in tumor cells
EMA2	Positive
EMA3	Negative in tumor cells
EMA4	Positive
EMA5	Negative in tumor cells
EMA6	Negative in tumor cells
EMA7	Negative in tumor cells
EMA8	Negative in tumor cells
EMA9	Negative in tumor cells
EMA10	Negative in tumor cells
EMA11	Negative in tumor cells
EMA12	Negative in tumor cells
EMA13	Negative in tumor cells
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EMA93	Negative in tumor cells
EMA94	Negative in tumor cells
EMA95	Negative in tumor cells
EMA96	Negative in tumor cells
EMA97	Negative in tumor cells
EMA98	Negative in tumor cells
EMA99	Negative in tumor cells
EMA100	Negative in tumor cells

IMPRESSION:
 IHC profile support the diagnosis of Wilms' tumor.

Figure 6: Immunohistochemistry of the tumor

3. Review of literature

Extra renal occurrence of Wilms' tumor was mentioned as early as 1961 by Moyson et al [4] as mediastinal Wilms' tumor and in 1979 by Roy G.K. et al [5] in retroperitoneal location. Since then there have been periodic reports of extrarenal Wilms' tumor located in various regions, other than retroperitoneum in mediastinum, inguinal region [6], sacrococcygeal area [7] and testis [8]. Wilms' tumors are thought to occur in developmentally aberrated metanephric blastema. It matures into normal renal tissue by 36 weeks of gestation. The metanephric blastema persisting after last period of gestation may be harbinger of Wilms' tumor [9]. The nephrogenic rests can be unifocal or diffuse and multifocal. The nephrogenic rests may consequentially undergo sclerosis, lie dormant, become hyperplastic or transform into malignancy [10]. The nephrogenic rests can be seen placed in ectopic locations like inguinal canal, lumbosacral area, adrenal glands, thorax, colon and heart [11]. They may be associated with some syndromes like Beckwith-Weidemann syndrome, hemihypertrophy or aniridia [12]. A small percentage about 1%, of nephrogenic rests undergo malignant transformation, and this is result of mutation of WT1 gene at 11p location [13]. Hence the tumors can be found occasionally outside kidneys in the locations of ectopic nephrogenic rests. Kim YW et al suggested possibility of pluripotent cells forming hybrid of two neoplasms with nephroblastomatous elements with teratomatous tissue leading to extrarenal Wilms' tumor [14]. Though ectopic nephrogenic rests have been diagnosed as early as 6 months, age of child at presentation with extrarenal Wilms' tumor doesn't differ much from the renal Wilms' tumor and is consistently reported between 2 to 5 years. A case of age of 52 yrs has also been reported as adult extrarenal Wilms' tumor [15]. The presentation of the tumors has

been noted as asymptomatic mass in the location of the primary. It has nonspecific symptoms of abdominal pain or discomfort, weight loss and appetite changes. The mass effect may resemble a hernia in inguinal region [16], vaginal bleed [17], testicular swelling [18], or chest symptoms associated with mediastinal mass. No associated pediatric anomalies like Beckwith-Weidemann syndrome or congenital defects related to spine, cardiovascular system, gastrointestinal and genitalia were reported by any of the above authors. The diagnosis of extra renal Wilms' tumor should mean nephroblastoma in extrarenal location that is not a metastasis, there should be no primary tumor in kidneys and kidneys should not be supernumerary [19]. Extrarenal Wilms' tumors are mentioned in literature as teratoid or classical Wilms tumor. They usually have favourable histology. There is no defined staging system for extrarenal Wilms' tumor and National Wilms' Tumor Study Group (NWTSG) classifies all extrarenal Wilms' tumors as stage II or above [20]. Haiyan Liang et al found only 3% of extra renal Wilms' tumor cases to be metastatic. Older age, larger tumor size and preoperative tumor rupture were risk factors for recurrence and metastases in their study [21]. Immunohistochemistry can help in aiding diagnosis of Wilms tumor in extra renal location. Blastemal elements express CD56, CK22 and CK 8, while epithelial cells immunostain with CK22, CK18, CD57. Stromal cells express SMA, Actin Desmin, CD 34 [22]. Immunostaining with WT1 occurs in epithelial and blastemal components and helps differentiating Wilms' tumor from other pediatric tumors [23]. The investigations and treatment of extrarenal Wilms' tumor doesn't differ from renal Wilms' tumor. The NWTSG or Societe Internationale D'oncologie Pediatrique (SIOP) don't suggest a biopsy preoperatively or before neo-adjuvant chemotherapy for localised tumor. A biopsy is suggested before

chemotherapy if the tumor is inoperable or metastatic. The NWTs suggests upfront surgery followed by chemotherapy while SIOP suggests neo adjuvant chemotherapy followed by surgery [24]. National Comprehensive Cancer Network advises upfront surgery for most of the cases as they are unilateral and operable. Those tumors that are deemed inoperable or metastatic or are bilateral should be offered neoadjuvant chemotherapy [25]. In our case we did a preoperative biopsy as hepatoblastoma was the primary diagnosis. The location of tumor was left lobe of liver with a normal α -fetoprotein value. The diagnosis was made on preoperative biopsy and confirmed with Immunohistochemistry. Since the tumor was over 20 centimeters in size, we opted for neoadjuvant chemotherapy followed by surgery. We performed a left hepatic lobectomy as left lobe was present as very thin rim of normal liver parenchyma. The post operative histopathology confirmed Triphasic Wilms tumor.

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