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CASE REPORT

Wilms' tumor may be a rare presentation of ovarian neoplasms in girls

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ABSTRACT

Extrarenal Wilms' tumor is a rare malignant disorder. It represents 0.5-1% of Wilms' tumor. Management for ERWT is challenging regarding its pathological diagnosis, staging, treatment and prognosis. Here we report a 4 years old girl, presented with huge pelvi abdominal mass originating from the left ovary which was seen by radiologic assessment as a large lobulated heterogeneously enhancing mixed solid and cystic retroperitoneal mass away from both kidneys, occupying most of the abdomen with pelvic extension; it was displacing bowel loops and pancreas, measuring 13 x 15 x 16 cm without evidence of enlarged lymphadenopathy. The mass was diagnosed postoperatively as an extrarenal Wilms' tumor proved by immunohistochemical staining for Wilms' Tumor1 (WT1) with the negative reaction for other markers that exclude other diagnostic possibilities. The patient had an aggressive presentation and advanced disease. The diagnosis was difficult due to the lack of specific radiologic stigmata and aberrant origin. She was treated with aggressive chemotherapy to suit the advanced stage of her disease and unfavorable histology pattern.

Keywords: ovary, Wilms.

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INTRODUCTION

Although Wilms' tumor is a common pediatric malignancy and the most common renal tumor during childhood, extrarenal Wilms' tumor (ERWT) is a rare tumor representing only 0.5-1% of Wilms' tumor cases (Alexander et al., 2017; Cao et al., 2017). Management for ERWT is challenging regarding its pathological diagnosis, staging, treatment and prognosis (Shojaeian et al., 2016). Here we report an ERWT tumor originating from the left ovary in a 4-year-old girl with unfavorable histology and aggressive disease. She was treated surgically and with adjuvant chemotherapy and radiotherapy of Wilms' tumor with a good initial response.

CASE DESCRIPTION

A 4-year-old girl suffered from progressive abdominal enlargement and constipation for 2 weeks. On examination, a hard, non-tender, ovoid huge pelvic abdominal mass extending above the umbilicus was palpable. Abdominal and pelvic ultrasonography showed huge partially cystic, partially solid mass occupying most of the abdomen and pelvis. Post-contrast Computed tomography (CT) scan revealed a large lobulated heterogeneously enhancing mixed solid and cystic retroperitoneal mass away from both kidneys, occupying most of the abdomen with pelvic extension; it was displacing bowel loops and pancreas, measuring 13 x 15 x 16 cm without evidence of enlarged lymphadenopathy (Figure 1).

Laboratory investigations (full blood count, ESR and LDH) were all unremarkable. Also, tumor markers [αfetoprotein (AFP) and βhuman chorionic gonadotropin (βHCG)] were within normal values. The patient condition

progressed to severe abdominal pain, absolute constipation with persistent vomiting and it was diagnosed as an acute intestinal obstruction that necessitated urgent surgical exploration. The abdomen was entered through an upper abdominal transverse transperitoneal approach. On exploring the abdomen hemorrhagic ascites was detected and aspirated. The left ovary was the site of a large dumbbell-shaped mass approximately 15x20x15 cm. The mass had a whitish_smooth medial surface while the lateral aspect showed a rough cauliflower-like tumor which appeared to be an extension of the mass through the ovarian capsule (Figure 2). Multiple hugely enlarged bilateral internal and common iliac, aortocaval and para-aortic lymph nodes were seen in addition to many omental nodules. The left ovary and fallopian tube were excised in addition to total omentectomy and radical lymphadenectomy extending up to the level of the renal veins. Sampling of the mesenteric lymph nodes was done in addition to an appendectomy. All these were sent for histopathological examination.

Surprisingly, the tumor showed triphasic histology (blastemal, epithelial, and stromal components) which is characteristic of Wilms' tumor (Figure 3). The blastemal element consisted of highly cellular sheets of primitive small blue cells with scant cytoplasm. There were frequent foci that show large hyperchromatic nuclei and frequent atypical mitotic figures (diffuse anaplasia) (Figure 4). The epithelial components were in the form of abortive glomeruli and tubules while the stromal portion was composed of fibroblast-like cells with oval nuclei. Immunohistochemical (IHC) staining showed diffuse positive membranous reaction for pan cytokeratin (AE1/AE3), diffuse positive nuclear reaction for WT1 (Figure 5A), high expression of ki67 (Figure 5B), focal positive membranous reaction for CD56. While negative staining for Placental Alkaline Phosphatase (PLAP) (Figure 5C), AFP, Glypican 3, cytokeratin7, HCG, CD30 & CD117. Specimens from para-aortic, left and right common iliac, aortocaval lymph nodes as well as omental nodules all were infiltrated by the same tumor tissue.

The post-operative period was uneventful and 2 weeks later she was referred to the pediatric oncology unit for adjuvant therapy. C-T scan for the chest and abdomen was performed for the patient. It revealed multiple scattered subpleural pulmonary nodules in both lung fields and enlarged internal and external iliac lymph nodes (Figure 6). Management of the patient was based upon the Children Oncology Group study which recommended further augmentation of therapy for stages II to IV diffuse anaplastic tumors using a treatment regimen containing cyclophosphamide, carboplatin, and etoposide alternating with vincristine, doxorubicin and cyclophosphamide (regimen UH-1). As she was categorized to have an advanced stage of the disease (stage IV) with unfavorable histology (diffuse anaplasia). Radiotherapy was planned to be postponed till assessing the response to chemotherapy after 6 weeks. Six weeks later, the patient was evaluated for chemotherapy response which was marvelous with the complete disappearance of the pulmonary metastases (Figure 7).

The patient was referred to receive radiotherapy for the abdomen concomitantly with chemotherapy. Radiotherapy was given as whole abdomen external beam radiation therapy using 6-MV linear accelerator (ELECTA) after CT simulation and 3-dimensional conformal radiotherapy planning. AP/PA beams and segments arrangement with multileaf collimator (MLC) shaping are produced. The upper margin of the abdominal field included the diaphragm down to the lower border of the obturator foramen ad pelvic floor. The acetabulum and femoral head were excluded from the irradiated volume. The outline of the organs at risk was delineated with respect to dose constraints. Gross target volume is the tumor volume before surgery using both the available CT scan before surgery and operative details with involved omental nodules and lymph nodes shown in the pathological report. Planning target volume (PTV) coverage goals were 100% of PTV receives 95% or more of prescription dose and the maximum dose is less than 107% of the prescription dose.

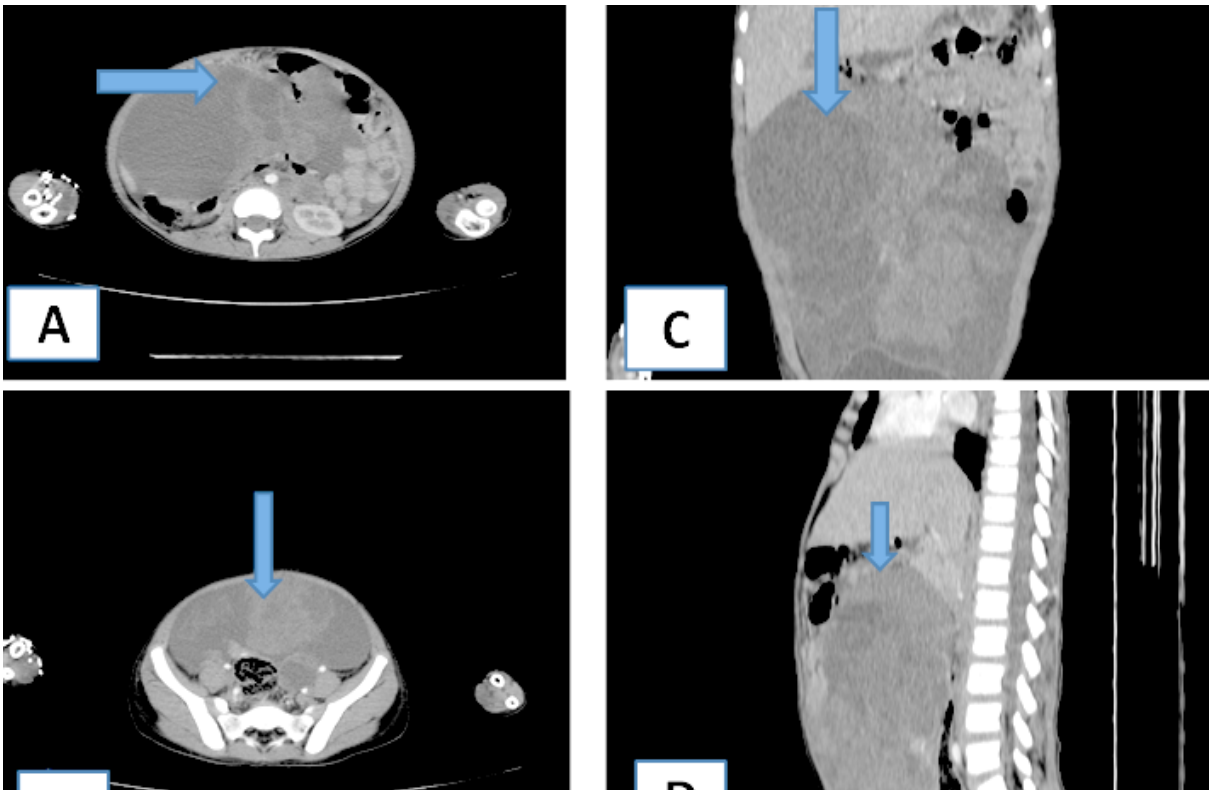

Figure 1. Post-contrast abdominal and pelvic C-T scan of 4 years old girl with palpable pelvi-abdominal mass. It shows huge abdominal heterogeneously enhancing mass with pelvic extension measuring 13 x 15 x 16cm: A Axial view of the huge mass occupying abdomen, B Axial view of the pelvic part of the mass, C; coronal view showing the extension of the tumor from the pelvis to the upper abdomen, D; sagittal view revealing of the tumor.

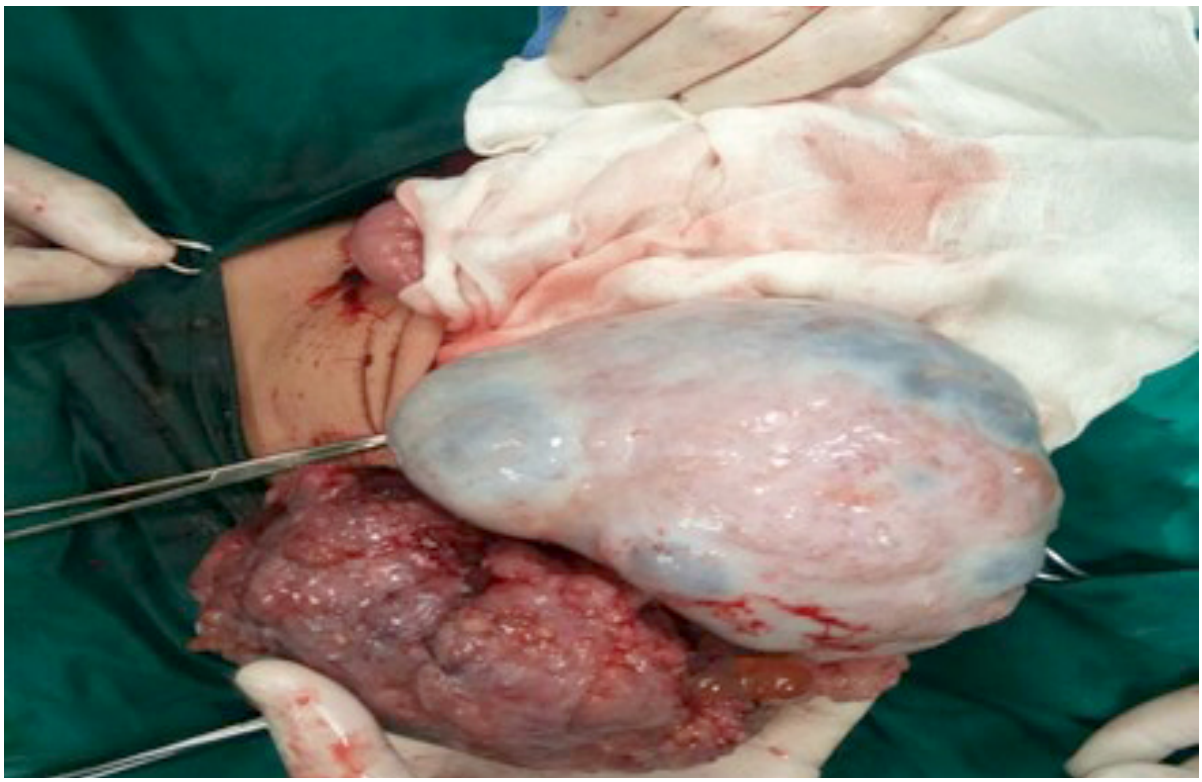

Figure 2. Intraoperative photo showing: large dumble shaped mass originating from the left ovary consisting of whitish smoothly surfaced part and another hard, rough cauliflower part.

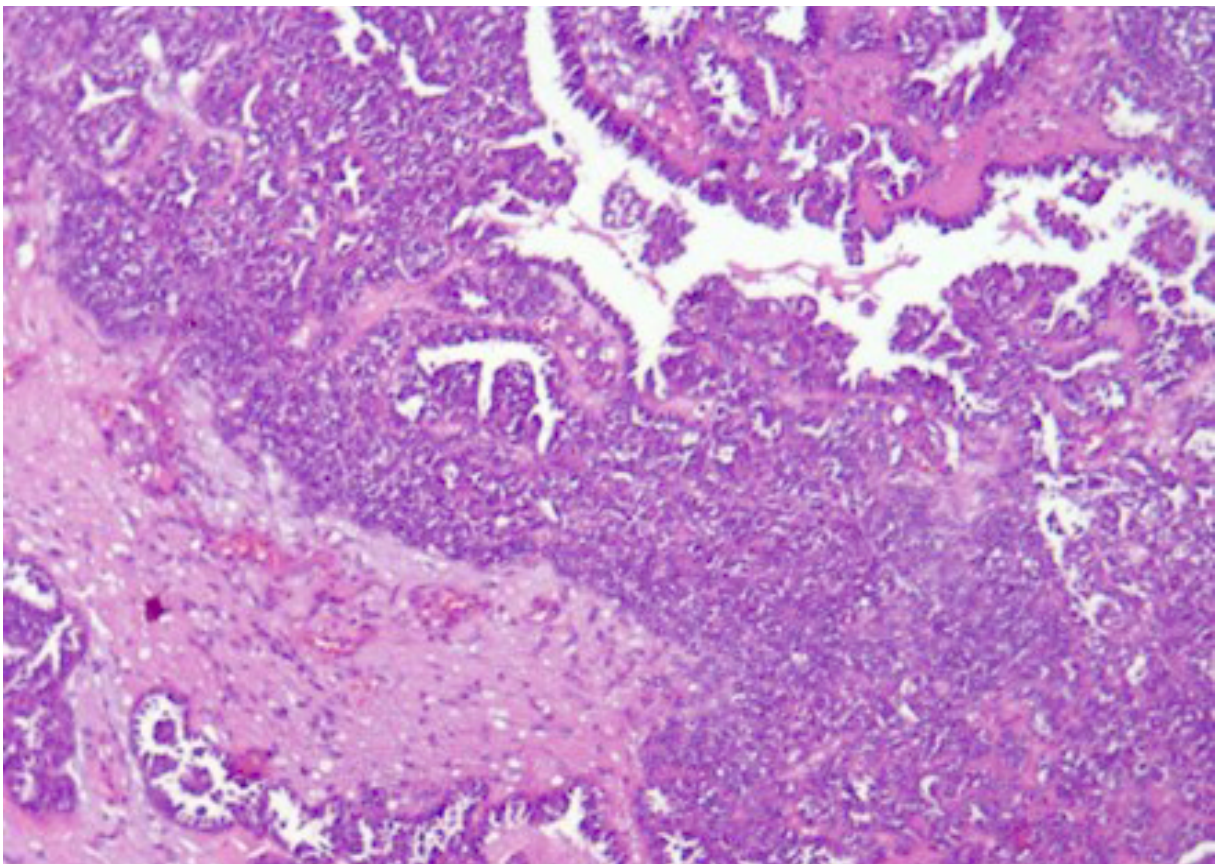

Figure 3. Photomicrographs showing: triphasic elements of Wilms' tumor, consisting of blastemal, epithelial and stromal elements (x200original magnification H&E)**.**

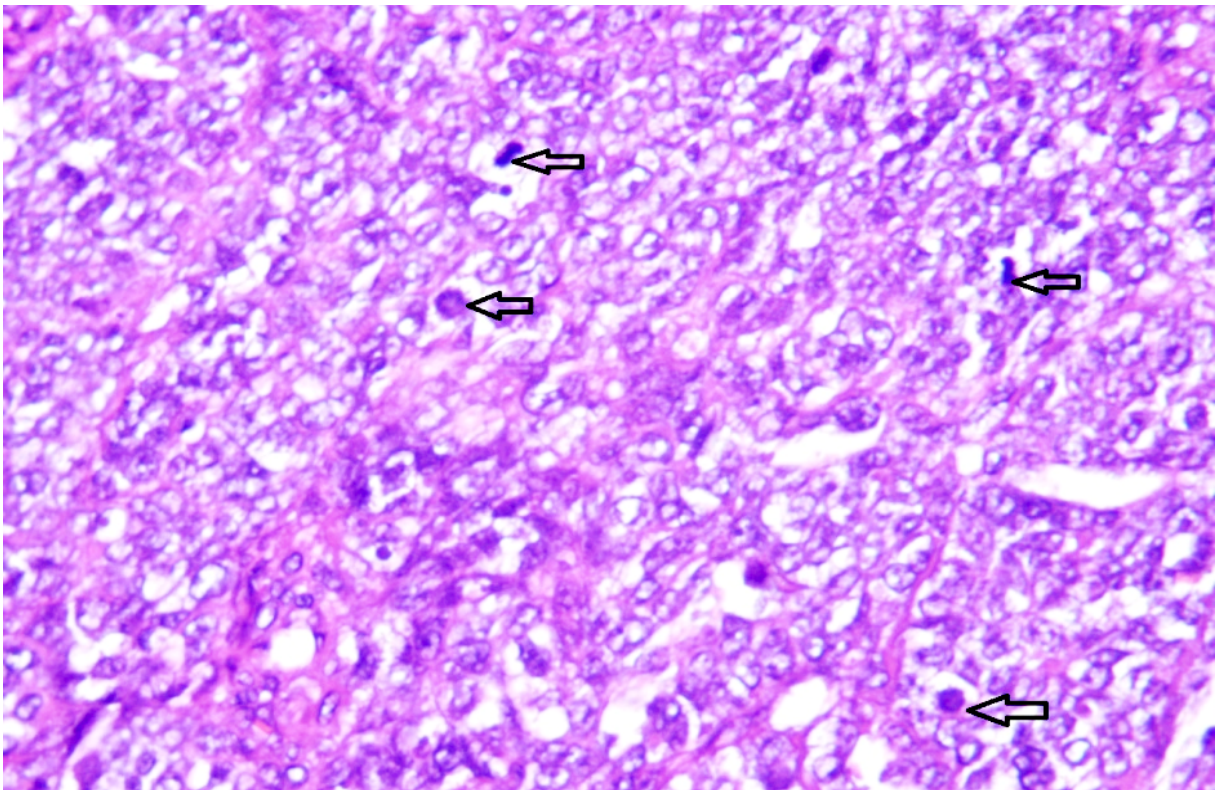

Figure 4. Photomicrographs showing anaplastic features (frequent atypical mitotic figures; arrows) (x200 original magnification H&E)**.**

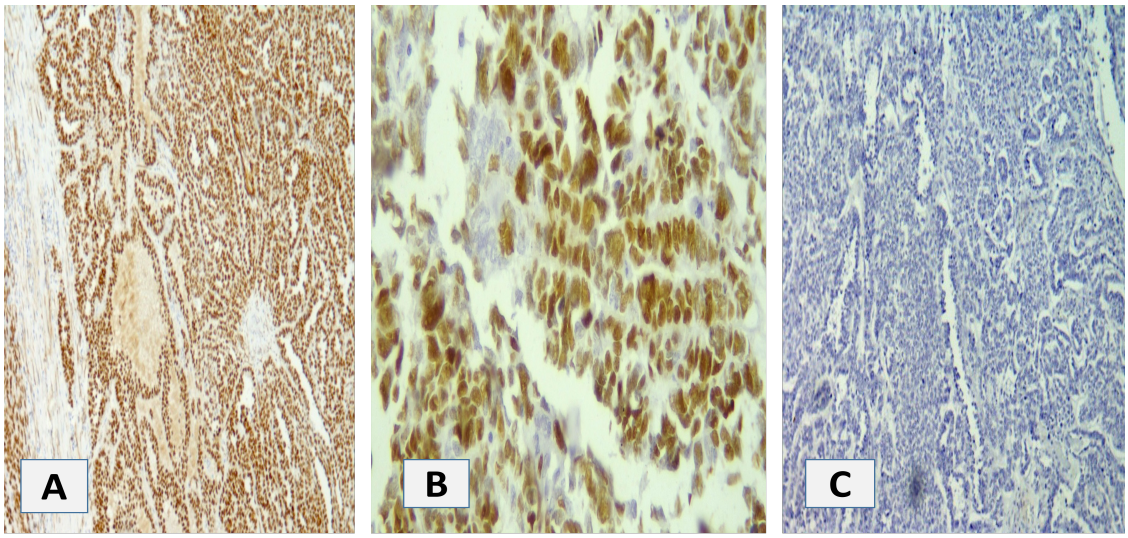

Figure 5. Immunohistochemical profile of the tumor (A), Photomicrograph showing the diffuse positive nuclear reaction of tumor cells for WT1 (X100 WT1); (B), Photomicrograph showing: high nuclear reaction of tumor cells for ki67 (X200 WT1); (C), Photomicrograph showing the negative reaction of tumor cells for PLAP (x100 PLAP).

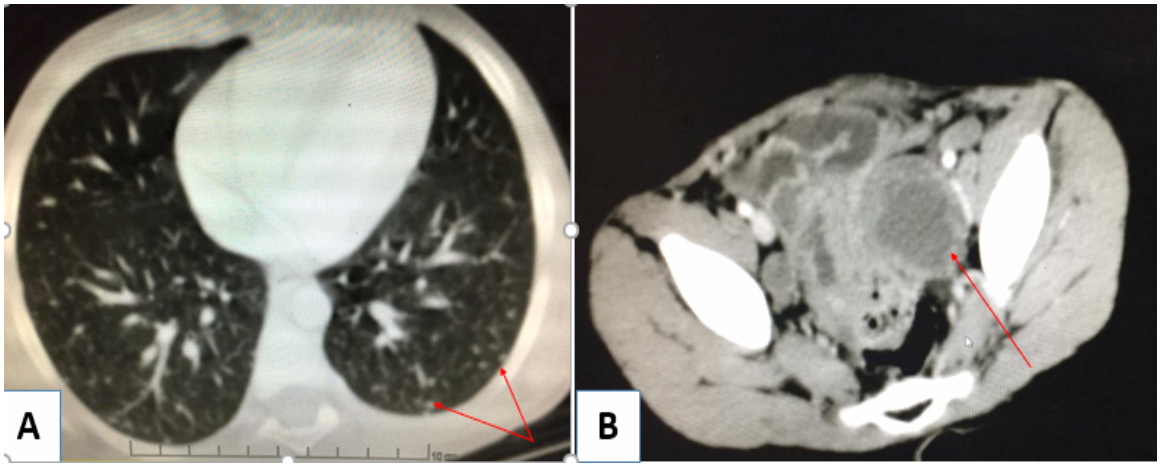

Figure 6. CT chest showing multiple small subpleural pulmonary nodules (A), CT pelvis showing multiple external and internal iliac Lymph nodes, the red arrow points to a large metastatic lymph node with central necrosis (B).

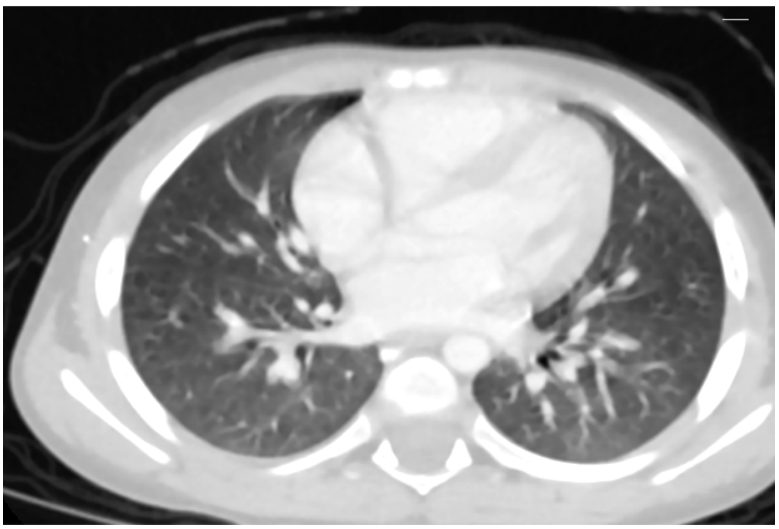

Figure 7. CT chest showing complete disappearance of the pulmonary nodules after 6 weeks of chemotherapy.

Treatment was given in once-daily fractionation regimen of 10.5 Gy in a 7 fractions of 1.5 Gy per fraction, 5 days per week concurrent with weekly vincristine. Four months later, the patient is still completing her chemotherapy regimen and she is in remission.

DISCUSSION

Extrarenal Wilms' tumor is an exceptionally rare neoplasm of children (Armanda et al., 2012). However, there are few reported cases in adults (Alexander et al., 2017). The exact origin of the extrarenal tumor is still unclear, however, it is believed to originate from ectopic nephrogenic rests (Cooke et al., 2009). including ectopic metanephric blastema (Broecker et al., 1989), mesonephric duct remnants (Fernandes et al., 1989) or malignant transformation of cells with persistent embryonal potential (Andrews et al., 1992).

Different locations were described for this tumor including the inguinal area, uterus, cervix, vagina, ovary, testis, bladder, sacrococcygeal and lumbosacral regions, retroperitoneum, and thorax (Arkovitz et al., 1996; Arda et al., 2001; Deshpande et al., 2002; Oner et al., 2002; Houben et al., 2007; Song et al., 2010; Taguchi et al., 2010) .

Since the tumor originated from the ovary, ovarian germ cell tumor was the first possible diagnosis albeit it was not supported by elevated tumor markers (AFP or βHCG), furthermore histopathological examination was against such diagnosis (negative immunohistochemical reaction for PLAP, CD117, CD30, AFP and glypican 3). Also, the lack of any teratomatous components excludes teratoma with renal tissue. Diagnosis of Wilms' tumor was based upon its characteristic triphasic histology formed of blastemal, epithelial, and primitive stromal elements along with its immunohistochemical profile. Unlike most reported cases showing favorable histology patterns of the tumor (Shojaeian et al., 2016), unfortunately, the histology was unfavorable evidenced by the presence of diffuse anaplasia.

Primary extrarenal Wilms tumor of gynecologic tract is extremely rare with scattered case reports occurring in the ovary, uterus, corpus, and cervix. Only 9 cases of primary ovarian Wilms tumor have been reported to date (Turashvili et al., 2020). The diagnosis of ERWT is a real challenge as it lacks pathognomonic radiologic stigmata making the diagnosis is often established postoperatively (Shojaeian et al., 2016). National Wilms' tumor study (NWTS) staging system is usually applied for ERWT. Being beyond the renal capsule, there should not be stage I ERWT (Rojas et al., 2013). In the current case, stage IV was considered based on the presence of pulmonary metastasis. Treatment of ERWT is similar to intrarenal Wilms' tumor with complete surgical excision remains the key step in the treatment of ERWT followed by adjuvant therapy which is indicated for all cases (Morandi et al., 2013). This case was planned for intensive treatment consisting of 5 chemotherapeutic agents because of disseminated disease (stage IV) besides unfavorable tumor histology. Radiotherapy was delayed till evaluating the response to 6 weeks of postoperative chemotherapy to avoid total lung radiotherapy and whole abdominal irradiation was given. Similar to intrarenal Wilms', the prognosis of ERWT is based on histology and staging according to NWTS group (Armanda et al., 2012).

Though it is rare, ERWT should be considered in the differential diagnosis of retroperitoneal or inguinal masses in childhood. Diagnosis is challenging as it is often made after surgical removal. Staging and management follow the same principles of that of intrarenal Wilms' tumor. The Pediatric Oncology team should be aware of this rare tumor to manage properly.

ETHICAL CONSIDERATION

An informed consent was obtained from the patient's parent attending with her for publishing her daughter's clinical, surgical, and radiological data.

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CONFLICTS OF INTEREST

Authors declare no conflicts of interest.

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