Adult Extrarenal Teratoid Wilms’ Tumor: A Case Report

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Adult extrarenal teratoid Wilms’ tumor: a case report
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Abstract:
Background: A teratoid Wilms’ tumor (TWT) is a very rare histologic variant of the classical Wilms’ tumor. This tumor development outside kidneys is rare. Extrarenal TWT mainly occurs in children, and reports of adult-related cases are rare. Till now, only seven cases of extrarenal TWT have been reported, among which five cases occurred in children and another two cases was reported in adult.

Methods: A retrospective analysis of the clinicopathological characteristics of a case of extrarenal teratoid Wilms’ tumors (TWT) admitted to our hospital and analysis and summary with literature.

Results: The case was a 19-year-old woman that presenting with lower, painless abdominal discomfort, and an imaging of an ultrasound and MRI both showed a cystic-solid mass on the right ovary. The patient subsequently underwent a laparoscopic right ovarian cystectomy, The specimen is sent for pathological examination, microscopic examination showed typical morphological features of Wilms’ tumor and predominance of teratoid elements which comprising more than 50% of the tumor. The diagnosis was made as extrarenal TWT in right ovary. After 3 courses of chemotherapy, the patient remains without evidence of disease after twenty-six months of follow-up.

Conclusion: Our case is the third reported case of adult extrarenal TWT occurring in ovarian. Careful histomorphological examination and extensive immunohistochemical studies will allow for accurate diagnosis, We reported this case in order to improve the clinicians and pathologist’s understanding of tumors.

Key Words: extrarenal, teratoid Wilms’ tumor, ovary

Background
Wilms’ Tumor is the commonest malignancy of genitourinary tracts in children, A teratoid Wilms’ tumor (TWT) is a very rare histologic variant of the classical Wilms’ tumor with predominantly teratoma-like heterologous elements and the heterologous tissues account for more than 50% of the tumor [1]. This tumor development outside kidneys is rare. Extrarenal TWT mainly occurs in children, and reports of adult-related cases are rare. The treatment of TWT is the same as Wilms’ tumor. To the best of our knowledge, only seven cases of extrarenal TWT have been reported, among which five cases occurred in children and another two cases was reported in adult. Here, we reported an adult ovarian extrarenal TWT, exploring the clinicopathological characteristics of the tumor to improve the pathologist’s understanding of tumors.
Methods
A retrospective analysis of the clinicopathological characteristics of a case of extrarenal teratoid Wilms’ tumors (TWT) admitted to our hospital. The patient was a 19-year-old woman. She experienced lower, painless abdominal discomfort for three days. An ultrasound showed a cystic-solid mass measuring 15.9 cm on the right ovary. MRI showed there was round masses with long/T1 and long/short T2 mixed signal in pelvic cavity and the lower abdomen. The mass was measured 13.0X12.6X16.0cm, and it could see the T1 and T2 sensations separate the mass inside. (Fig. 1). The shape was irregular in right ovary. There was no evidence of extra-ovarian disease. The diagnose was concerning for ovarian teratoma, but other germ cell tumors were not excluded.

FIG. 1 MRI findings: there are a round mass with long/1 and long/short T2 mixed signal in pelvic cavity and the lower abdomen, and it can see the T1 and T2 sensations separate the mass inside. (A)Transverse plane and (B)Sagittal plane.

The levels of serous markers were measured; CA125 level were recorded as 81.55U/ml (normal levels are 0.1-35 U/ml), CA19-9 level were recorded 618.3U/ml (normal levels are 0.1-27U/ml). The remaining 3 test items: carcinoembryonic antigen (CEA), alpha-fetoprotein (AFP) and CA153 were all in normal level. Sex hormone test items were all in normal level, too, including Follicle stimulating hormone (FSH), luteinizing hormone (LH), estradiol (E2), progesterone (P), and testosterone (T).

Result:
The patient underwent a laparoscopic right ovarian cystectomy. The specimen is sent for pathological examination. Macroscopically, the mass of the right ovary measured 14X13X5cm and it was composed of a cystic and a solid part. The cyst wall of the tumor was ruptured and there was no fluid left inside the mass. The solid part was multinodular, fibrotic appearance with necrosis. It measured 13X13X5cm, and the cut surface was yellowish tan. The tumor was tender, and it displayed bone-like texture as well as grease in different areas (Fig. 2).
FIG. 2 Macroscopic findings, the cut surface had a yellowish tan, multinodular, fibrotic appearance with necrosis.

There were the primitive tubules in a pseudorosette formation (Fig. 3) beside teratoma-like elements. Frozen diagnosis was made malignant immature teratoma of right ovary. After the specimens are fixed with 10% neutral formalin, they are routinely fully collected and embedded into slices. A microscopic examination sections from the solid mass showed mainly mature teratomatous elements along with classical triphasic elements of Wilms’ tumor (Fig. 4).
FIG. 3 Microscopic findings in frozen section see the primitive tubules in a pseudorosette formation.
The components of classic Wilms tumor include: 1. Embryonic undifferentiated embryo components: cells are small round and oval, nuclei are deeply stained, nucleoli are not obvious, nuclei are overlapped, mitotic activity is active, and cytoplasm is minimal. 2. Primitive mesenchymal components: cell spindle or fiber cell-like structure. 3. The structure of embryonic renal tubules, with primitive chrysanthemum-shaped clusters, glandular tube-like and ribbon-like structures (Fig. 5). Wilms tumor component accounts for 20% of the mass.

The other of the trademark features of extrarenal Wilms’ tumor (TWT) is predominantly teratoma-like heterologous elements including: stratified squamous epithelium, mucous columnar epithelium, ciliated columnar epithelium, and sebaceous glands derived from endoderm; well differentiated
adipose tissue, fibrous tissue and cartilage derived from mesoderm (Fig. 6). Heterogeneous ingredients account for about 80% of the mass.

FIG. 6 Microscopic findings teratoma-like heterologous elements, including stratified (A)squamous epithelium, (B)mucous columnar epithelium and breast glands, and (C)well differentiated adipose tissue, fibrous tissue and cartilage (X20. Hematoxylin and eosin staining).

Immunohistochemical examination showed that embryonic kidney germs and tubule-like structures expressed Wilms’ tumour gene(WT1) and SALL4 (Fig. 7a,b); embryonic kidney germs, renal tubules and primitive mesenchymal components expressed CD56(Fig. 7c). Primitive mesenchymal components expressed Vimentin. Few embryonic renal tubules expressed cytookeratin (CK)-pan and epithelial membrane antigen(EMA). The Ki-67 proliferation index of tumor cells was 60%. The components of embryonic kidney germ, renal tubules and primitive mesenchyme was negative to AACT, carcinoembryonic antigen (CEA), alpha-fetoprotein(AFP), CD99, MyoD1, Myoglobin, Myogenin, smooth muscle actin(SMA), Caldeomon, Demin, S100, synaptophysin(Syn), chromogranin(CgA), Neuron specific enolase (NSE) and neurofilament protein(NF).

FIG. 7 Immunohistochemistry: embryonic kidney germs and tubule-like structures displays both WT1 (A; magnification X20) and SALL4 positivity (B; magnification X20). Embryonic kidney germs, renal tubules and primitive mesenchymal components displays CD56 positivity (C; magnification X20).

A diagnosis was made by two experts in gynecologic pathology through pathology remote consultation: extrarenal TWT in right ovary.

The patient then underwent 3 courses of chemotherapy (cisplatin, etoposide and bleomycin), and she remains without evidence of disease after twenty-six months of follow-up.

Discussion
Wilms’ Tumor is the commonest malignancy of genitourinary tracts in children and it may manifest genetic abnormalities in one of the two regions in the short arm of chromosome 11 namely 11p13 (WT1) and 11p15 (WT2) [2]. The term “teratoid Wilms’ tumor” was first used in
1984 to describe a Wilms tumor with extensive heterologous elements. The heterologous elements should comprise more than 50% of the tumor in an organoid arrangement [1]. Diagnosis of extrarenal Wilms’ Tumor (EWT) must exclude a primary tumor in kidneys. Development of Wilms’ tumor outside kidneys is rare and has been reported to occur in a variety of locations, including the sacrococcygeal region, adjacent to kidneys and uterus. There are two types of EWT: (1) EWTs found as components of teratomas (teratoid WT) and (2) pure EWTs of one tissue type. Teratoid EWT are rarer than pure EWTs[3, 4].

Extrarenal teratoid Wilms’ tumor (TWT) is a rare histologic variant of the classical Wilms’ tumor, containing predominantly heterologous tissues [5]. Immunohistochemical of Wilms’ tumor displayed positive for WT1, CD56, CD99, SALL4 and Neuron-Specific Enolase (NSE), and negative for ER, PR, CD10 and GFAP [6] [7] [8]. Extrarenal Teratoid Wilms tumors are rare with approximately 7 reported cases to date. Only 2 cases were occurred in adult and both tumor were located in ovary. The occurrence sites include retroperitoneum, pelvic cavity (uterine-rectal space), sacrococcygeal, vagina, ovary and ureteropelvic. Among these cases, the youngest is 1 day, the oldest is 36 years old. There were 3 males and 4 females [3, 5, 8-11]. The tumor may accompany with congenital dysplasia. Baskaran reported a TWT was in association with horseshoe kidney in a 3-year-old boy[5]. Few cases were associated with elevated serum AFP [10, 12]. The extrarenal TWT has led to the debate on whether the origin is embryonic or neoplastic. Some authors thought the tumor originated from teratoma, and they preferred to call the tumor as a teratoma with a predominance of nephroblastic elements rather than that of an extrarenal teratoid Wilms’ tumor[10]. But most authors believed that the origin of this tumor was the embryonic remnants of mesonephric tissue [3].

The differential diagnosis of an extrarenal (ovarian) TWT includes metastatic Wilms’ tumor, immature teratoma, malignant mixed Müllerian tumor (MMMT) and neuroblastoma. The diagnosis of a metastatic Wilms’ tumor must exclude that there wasn’t a classic Wilms’ tumor in the kidneys. Immature teratoma was seen the embryonic immature and mature tissues are mixed which are from the three germ layers. Immature embryonic tissue is often neuroectoderm chrysanthemum or primitive neural tube tissue [13]. MMMT is usually presented in older postmenopausal patients. The tumor exhibits an admixture of malignant epithelial and mesenchymal tissue. Immunohistochemical markers PLPA, α-inhibin, HCG, AFP, CD30 are often positively expressed. Neuroblastoma was composed of diffuse small round or oval cells. The cells are relatively uniform and more frequent mitosis. Homer-Wright chrysanthemum cluster structure is common. Immunohistochemical markers were positive for CgA, Syn, NSE, and negative for WT-1 [14]. It is worth noting that most of these tumors may contain heterologous elements, the presence of characteristic morphologic features and additional immunohistochemical analysis may be helpful for an accurate diagnosis.

In general, most extrarenal TWT have favorable histology, and there were no metastases or recurrences during long-term follow up [5, 10].

At present, there was no standard clinical staging and treatment plan for TWT, and the clinical staging and treatment plan for Wilms tumor is still adopted. However, because teratoma-like Wilms tumor contains a large number of well-differentiated heterogeneous components and is not sensitive to radiotherapy and chemotherapy, some scholars believe that surgical removal of the tumor is the best treatment method [5], and better treatment methods need to be further improved.

**Conclusion**

In summary, the case is the third reported case of teratoid Wilms’ tumors in ovary. It can be
diagnostically challenging and requires exclusion of other entities. Careful histomorphological examination and extensive immunohistochemical studies will allow for accurate diagnosis.

**Declarations**

**Ethics approval and consent to participate**

Our study was approved by the ethical committee of Maternal and Child Health Hospital of Ganzhou City, Jiangxi Province, China.

**Consent for publication**

The authors agree to public in cancer cell international.

**Consent for publication**

The patient of case reports has consented for publication.

**Availability of data and materials**

Not applicable

**Competing interests**

The authors declare that they have no competing interests.

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**Authors’ contributions**

LL designed and performed data analysis and wrote the manuscript. CH contributed to interpret imaging data and revised the manuscript. Both authors read and approved the final manuscript.

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