

# Extrarenal Testicle Wilms Tumor Followed the Operation of Left Hernia Sac High Ligation and Right Cryptorchidism Descent Fixation: A Case Report

**Jinjing Wang**

the affiliated hospital of zunyi medical university

**Yao Li**

Fifth Affiliated Hospital of Zunyi Medical University

**Shuai Luo**

Affiliated Hospital of Zunyi Medical University

**Hong Zheng** (✉ [313571974@qq.com](mailto:313571974@qq.com))

affiliated hospital of zunyi medical university

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## Case Report

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# Abstract

**Background:**The extrarenal Wilms tumor is a rare malignancy in the pediatric population. So far, no extrarenal testicle Wilms tumor followed the operation of left hernia sac high ligation and right cryptorchidism descent fixation has been reported in the literature.

**Case presentation:** We researched an exceptionally unheard case of the extrarenal testicular Wilms tumor after the surgery of left hernia sac high ligation and right cryptorchidism descent fixation in a 2.8 years old boy. The patient was admitted to a hospital due to the evidence of huge scrotum mass six months after the operation of left hernia sac high ligation and right cryptorchidism descent fixation. A huge tumor ascending from the right testicular was found.

**Conclusions:** Histology examination revealed the typical triphasic Wilms tumor elements such as epithelial, mesenchymal, and blastemal areas. This study revealed histopathological, clinical, diagnostic, prognostic, and therapeutic characteristics of this infrequent tumor.

## Introduction

One of the utmost childhood solid malignancies, which results in almost 95% of renal malignancies in pediatrics is Wilms tumor. It usually occurs between the ages of 1 and 3 years [1]. The extrarenal Wilms tumor (EWSRT) is an extremely uncommon but thought-provoking body [2]. The approximate prevalence of EWSRT is about 1% of all cases of Wilms tumor.[3] In several organs, EWRT may take place. The most common extrarenal sites are retroperitoneal, lumbosacral, pelvic, inguinal, female genital organs, and paratesticular [4]. So far, three cases of paratesticular ERWT have been reported [5]. No extrarenal testicle Wilms tumor followed the operation of left hernia sac high ligation and right cryptorchidism descent fixation has been reported in the literature [6]. The EWSRT lacks characteristic clinical manifestations, laboratory tests, and imaging indication and its diagnosis mainly depends on pathological examination. Herein, we describe a case of extrarenal testicle Wilms tumor, followed by the operation of left hernia sac high ligation and right cryptorchidism descent fixation. The case is described completely in terms of its clinical, histopathological, immunohistochemical characteristics, differential diagnosis. Furthermore, this rare tumor case is discussed with reference to the literature to understand the tumor and improve its diagnostic accuracy.

## Case Presentation

The patient was a 2.8 years old boy born in June 2017. Shortly after the birth, it was found that his right scrotum was empty. The condition did not change significantly as the child grew up, and no special treatment was performed outside the hospital.

Then, the patient was admitted to our hospital in July 2018, at the age of 13 months, and was found with an empty right scrotum, presenting mass in the right inguinal area. The physical examination revealed the bilateral asymmetry of the scrotum. The left testicle was located in the scrotum, measuring 3mm × 3mm

× 5 mm. The light transmission test (-). A round-shaped mass on the left inguinal area was observed having a size of about 2.5 cm in diameter. The mass often came out to fall into the scrotum when the patient was crying, however, it could return to the abdominal cavity when the patient became quiet. No skin redness or inflammation/swelling was found during or after the situation. There was a palpable testicular like tissue in the middle part of the right groin region, measuring 2mm × 2mm × 3 mm in size. The performed ultrasonography revealed the intestinal loops in the left hernial sac without incarceration. The diagnosis of left ventral abdominal hernia without mechanical ileus accompanied with undescended testis in the right inguinal canal was made. The patient was discharged after the bilateral hernia sac high ligation and right testicular descent fixation surgery.

In March 2019 after eight months of surgery, the patient's parents found a painless mass in right scrotal, the mass had a rough surface and was the size of a soybean.

In September 2019 after fourteen months of surgery, the child was admitted to our hospital after when the mass of the right scrotal gradually increased to about the size of an egg. Upon physical examination, the bilateral groin area was found symmetrical and no mass was seen. The bilateral scrotum was asymmetrical; the right scrotum was significantly enlarged with a mass in the right scrotum. No abnormality was found in the left testicle. In the right scrotum, a heterogeneous dense mass with a definite boundary was exposed by Ultrasonography, measuring 36mm × 38mm × 40 mm, and with an increased blood flow in it. In the right testicle, a solid mass was exposed by Contrast-enhanced computed tomography, approximately 38mm × 40mm × 46 mm, and enhanced blood vessel shadows were seen. (Fig. 1). CT scan of the abdomen depicted no clue of distortion or parenchymal lesions of bilateral kidneys and both had a well-conserved morphology. There were no inflamed lymph nodes found. The rise of a malignant tumor from the right testis was the hypothesis. Laboratory analysis showed that serum  $\alpha$ -fetoprotein (AFP),  $\beta$ -subunit of human chorionic gonadotropin ( $\beta$ -HCG), neuron-specific enolase (NSE), lactate dehydrogenase, and carcinoembryonic antigen (CEA) were within the normal range.

Under general anesthesia, the patient received radical orchiectomy of the right testicular mass. The effective outcome was a distinct mass with a well-defined boundary and no invasion of the neighboring organ.

## **Pathological examination:**

Based on gross examination, it was found that the resected specimen was a solid mass measuring 38mm × 40mm × 46 mm, and was encapsulated by a fibrous capsule. The cut section showed a milky white cut-surface and the mass detached from the testis as well as the epididymis. After meticulous dissection of the tumor, the elements of the right testis were examined (Fig. 2).

Microscopic examination showed that the characteristics simultaneously exhibited a triphasic pattern of Wilms tumor (identical blastemal elements with attempted glomeruli and primitive tubules elements) with the adjacent foci of nephrogenic rests (Fig. 3). With the proof of immature testicular tissue in the peripheral area of the tumor, testicular tissue was just about entirely supplanted by neoplastic elements.

After adequate sampling and careful observation under the microscope, no teratoid element, renal carcinoma, anaplastic area, and tumor necrosis was detected in the entire specimen, and there were fewer mitotic figures.

Immunohistochemically, the stromal component was positive for vimentin and SMA. For WT1, CD56, CD99, the blastemal component was positive, while for epithelial membrane antigen (EMA), CK, PAX-8, and vimentin, the epithelial component was positive. The Ki-67 index in the blastemal component was about 80%, while the epithelial and mesenchymal approximately was 10%. NSE, S100, Inhibin, LCA, RCC, SALL4, P53, myogenin, SALL4, chromogranin A (CgA), Syn, S-100, and desmin were found negative (Fig. 4).

In conclusion, with the diagnosis of extrarenal testicle Wilms tumor, these outcomes were constant. The patient was generally in good condition after surgery and received 2 courses of postoperative chemotherapy using a regimen consisting of vincristine and actinomycin D in November 2019. Follow-up after one year of therapy follow-up, the patient is alive with no evidence of recurrence.

## Discussion And Conclusions

The Extrarenal Wilms tumor (EWSRT) is an extremely uncommon but challenging entity. It was first defined by Moyson et al. [7] in 1961. The most common extrarenal sites are retroperitoneal, lumbosacral, pelvic, inguinal, female genital organs, and paratesticular [4]. In the early years, Cryptorchidism is frequently known as a congenital deficiency [8]. It has been described that in the differentiation of structures involved in testicular descent, WT1 plays a direct role while genital abnormalities and cryptorchidism are frequently observed in male patients with germline WT1 mutations [8]. It is well documented that a tumor suppressor gene for Wilms' tumor is WT1 and accounts for 25% of ERWT cases [3]. So far, no extrarenal testicle Wilms' tumor followed by the operation of cryptorchidism descent fixation has been reported in the literature [6].

Currently, the exact mechanism of EWSRT which occurs in extrarenal tissue is unknown [9]. It is supposed that the intrarenal and extrarenal Wilms' tumors do not share the same embryological origin even having the same histology. Extrarenal Wilms' tumor may ascend from a more primitive mesonephric or pronephric origin and can have diverse positions, proposed by the generally acknowledged hypothesis for the pathogenesis of ERWT. While an embryonic neoplasm that stems from the metanephric blastema is broadly considered to be nephroblastoma [9]. During the embryologic growth period, the urogenital ridge and mesonephrons are in close proximity. Renal embryonic rests, metanephric blastema, or parts of the mesonephric duct might be replaced to the nearby gonad or Wolffian duct, which can continue in postnatal [9]. It is, therefore, expected that the ectopic metanephric blastemal cells in the paratesticular region may increase ERWT. The cases of ERWT found in the inguinal regions along the spermatic cord and testis, uterus, and sacrococcygeal region may be enlightened by the process [9].

EWSRT lacks characteristic clinical manifestations. The clinical presentation of ERWTs rests on the location and stage of the tumor [10]. So, its diagnosis is extremely difficult preoperatively and usually

occurs after surgery. In recognizing the characteristics of ERWT, imaging techniques, comprising the ultrasonography and contrast-enhanced computed tomography, may be beneficial. In some cases, additional imaging such as magnetic resonance study may be required [15]. An assured preoperative diagnosis of ERWT is impracticable despite these imaging tests. Currently, in an unknown childhood mass or ERWTs, the character of the intraoperative freezing segment is not reflected as a part of the surgical principle. The pillar of cure in most pediatric firm tumors is complete exclusion when valid. After the surgical resection of the tumor and pathological assessment of the sample, the analysis of ERWT is prepared almost at all times [11].

For pathological diagnosis of ERWT, the following four standards were suggested by Beckwith and Palmer: (1) extrarenal spot of primary neoplasm; (2) nascent blastematos spindle or round cell component; (3) abortive or embryonal tubular or glomeruloid structure; (4) no indication of teratoma or renal carcinoma [3]. In this case, microscopic examination showed the characteristically simultaneous exhibiting triphasic pattern of Wilms tumor (identical blastemal elements with attempted glomeruli and nascent tubules elements) with nearby foci of nephrogenic rests. With proof of normal testicular tissue in the peripheral area of the tumor, the testicular testis was juvenile, constant with the patient's age, and was almost totally supplanted by neoplastic elements. The analysis of teratoid Wilms tumor, which is a pretty diverse body in the pathogenesis and embryology with germ cell origin, is submitted due to the overabundance of heterotopic teratomatous elements exceeding 50% of the total microscopic field. We examined the whole specimen in multiple cuts to exclude the teratoid elements. In this case, through multiple materials and careful observation under the microscope, no teratoid element, renal carcinoma, anaplastic area, and tumor necrosis was found in the entire specimen.

The relationship and coexistence of ERWT with a horseshoe kidney have been described before in almost 13% of the testified ERWTs [12]. Both kidneys should be assessed specifically with a CT scan to eliminate any intrarenal tumor after pathologic validation of ERWT [3]. While encountering an abdominal mass in a patient with a horseshoe kidney, the examination of ERWT must be acknowledged [3]. In our case, the contrast-enhanced CT-scan indicated that the kidneys had well-kept morphology with no proof of deformity or parenchymal lesions.

Differential diagnosis comprises of major intrarenal Wilms tumor with extrarenal metastasis, malignant teratoma, neuroblastoma, embryonic rhabdomyosarcoma, and embryogenic germ cell-derived tumors.

(1) The diagnosis of extrarenal Wilms tumor first requires the exclusion of the intrarenal Wilms tumor to metastasize outside the kidney, which can be identified by imaging examination of bilateral renal space-occupying lesions. In this case, CT showed regular kidney morphology, and no obvious abnormal density was found in the parenchyma.

(2) Malignant teratoma: Immature teratoma shows mature and immature tissues in various germ layers. The diagnosis is based on the presence of neuroectodermal daisy clusters or primitive neural tubes, and both the mature and immature neural tissues with multiple germ layers mixed distribution. In our case, the three components of primitive germ component, epithelium-like component, and mesodermal

component had been found. The presence of teratomatous components was excluded by the extensive grossing of the variegated areas with detailed histological analysis, thereby it falls into the diagnosis of ERWT.

(3) Neuroblastoma: The tumor cells of neuroblastoma are primitive small cells with deep nucleus staining and inconspicuous nucleoli. ERWT is easily confused with neuroblastoma when it is dominated by primitive germ and lacks epithelial components. But Homer-Wright chrysanthemum is common in neuroblastoma. Immunohistochemically, neuroblastoma expresses cgA, Syn, and NSE, but does not express WT-1. In our case, the appearance of renal tubular structures with different degrees of differentiation in tumors, no Homer-Wright chrysanthemum was found. Immunohistochemical staining showed that the tumor cell is positive for WT-1 and negative for cgA, Syn, and NSE. All of these highly supports the diagnosis of ERWT and excludes neuroblastoma.

(4) Embryonic rhabdomyosarcoma: The tumor tissue is mostly primitive stellate or small round cells. Primitive mesenchymal components and rhabdomyoblasts of different stages are mixed in different proportions, but epithelial-like components do not appear. Immunohistochemical, staining showed that the tumor cells express desmin and myoglobin. In our case, the histological and immunohistochemical characteristics support the diagnosis of ERWT and exclude embryonic rhabdomyosarcoma.

(5) Embryogenic germ cell-derived tumors: These tumors are composed of multiple tumor components. Seventy-five percent of yolk sac tumors, seventy percent of embryonal carcinomas, and sixty-two percent of teratomas have a higher serum AFP level. However, in patients with Wilms' tumors, a boost of such protein is rarely found. In our case, laboratory analysis showed that serum AFP, CEA,  $\beta$ -HCG, and NSE were well within the normal range, which discriminated ERWT from embryogenic germ cell tumors, so it supports the diagnosis of ERWT.

No systematized standards have been explained so far to cure these lesions due to their uncommonness [13]. Surgical removal proves to be the main stride in the treatment of ERWT. It is broadly assumed that for ERWTs staging, National Wilms' Tumor Study (NWTS) system is likely to be very useful [14]. Despite suitable histopathology, for all ERWT cases, adjuvant chemotherapy is suggested postoperatively. Bearing in mind the NWTS protocols, the chemotherapy regimen is figured out by histology and phase of the tumor. For unresectable tumors and those with gross residue, recurrence, or metastasis, radiotherapy is suggested [3]. In this case, the child was younger and radiotherapy may have a greater impact on the growth and development of the child and the organs of the reproductive system. Therefore, only chemotherapy was preferred with the regime of vinpocetine and dextomycin. At present, two courses of chemotherapy have been completed, and the follow-up has been good so far, and no recurrence of the tumor has been seen.

## Conclusion

In summary, we reports the first case of extrarenal testicle Wilms tumor followed left hernia sac high ligation and right cryptorchidism descent fixation. We thoroughly described its clinical, histopathological,

immunohistochemical characteristics with differential diagnosis. Even though ERWT is rare in childhood, it should be considered as a differential diagnosis of retroperitoneal or inguinal masses. Awareness of ERWT improves diagnosis accuracy.

## Abbreviations

CT

computed tomography; EWSRT:extrarenal Wilms tumor; AFP:α-fetoprotein; β-HCG:β-subunit of human chorionic gonadotropin; NSE:neuron-specific enolase; CEA:carcinoembryonic antigen; CgA:chromogranin

A

## Declarations

### Ethics approval and consent to participate

This case report was approved by the Ethics Committee of the Affiliated Hospital of Zunyi Medical University. Written informed consent was obtained from the patient for publication of this clinical case report.

### Consent for publication

Written informed consent was obtained from the family of the patient for publication of this case report and any accompanying images.

### Availability of data and materials

All the data regarding the findings are available within the manuscript.

### Competing interests

The authors declare that they have no competing interests.

### Author details

Jinjing Wang, PhD: Department of Pathology, the First Affiliated Hospital of Zunyi Medical University, Guizhou, P.R. China.

Yao Li, BS: Department of Pathology, the First Affiliated Hospital of Zunyi Medical University, Guizhou, P.R. China.

Shuai Luo, BS: Department of Pathology, the First Affiliated Hospital of Zunyi Medical University, Guizhou, P.R. China.

Hong Zheng, MD: Department of Pathology, the First Affiliated Hospital of Zunyi Medical University, Guizhou, P.R. China.

### **Authors' contributions**

Resources: Shuai lu, Yao Li,.

Writing – original draft: Jinjing Wang.

Writing – review & editing: Jinjing Wang, Hong Zheng.

All the authors have read & approved the final manuscript.

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### **Conflict of Interests**

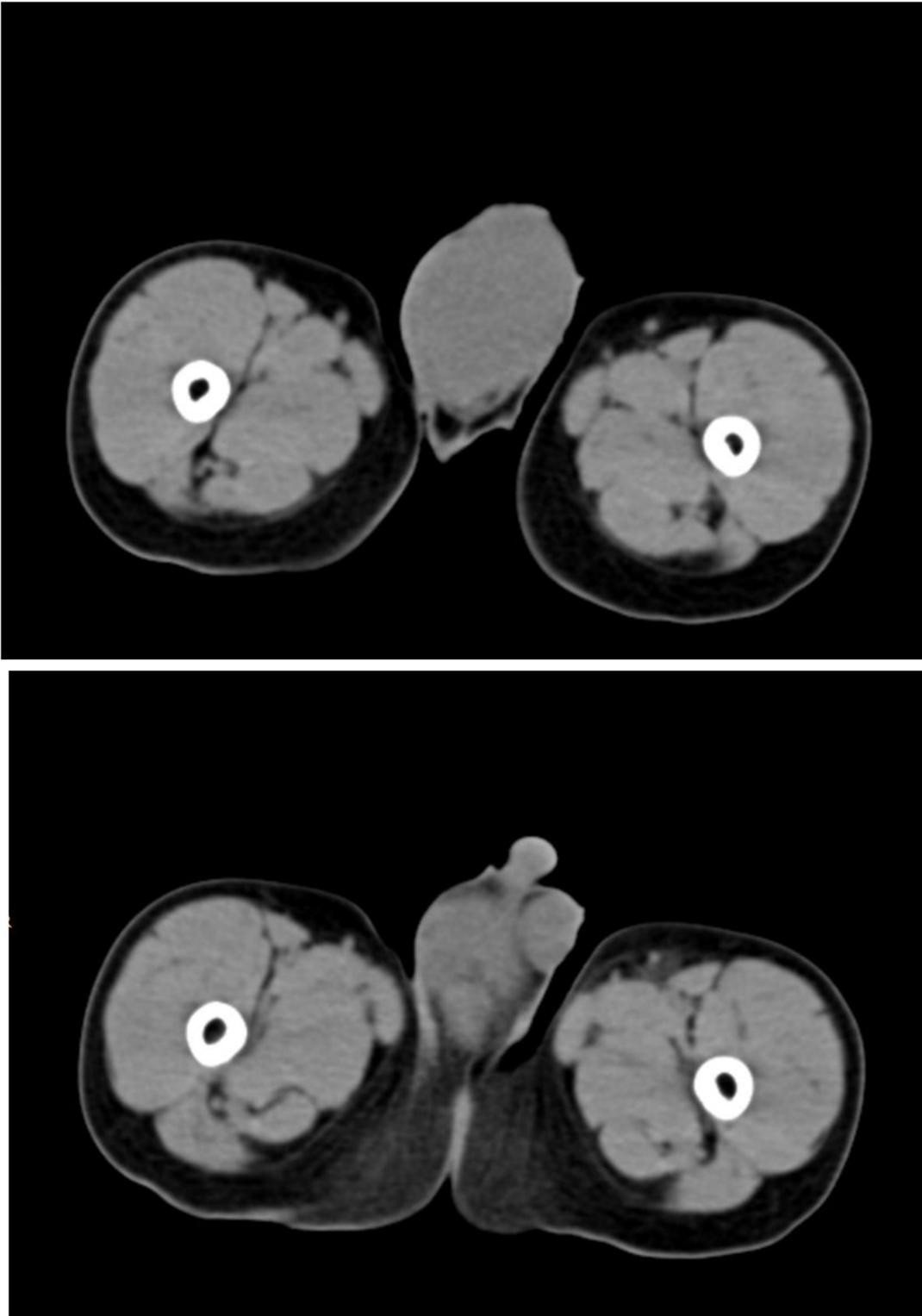
Regarding research, authorship, and/or publication of this article, the authors state no possible conflicts of interest.

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## Figures



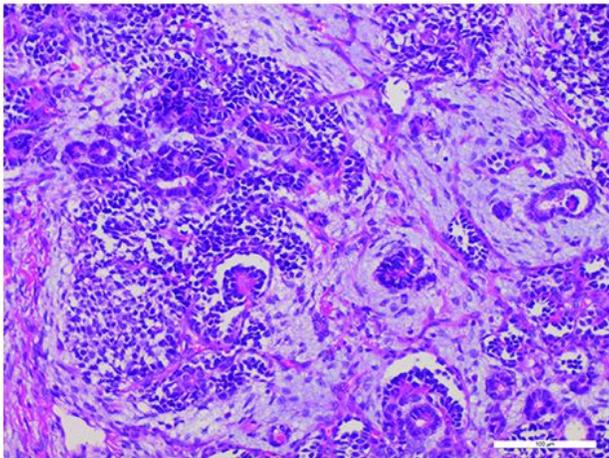
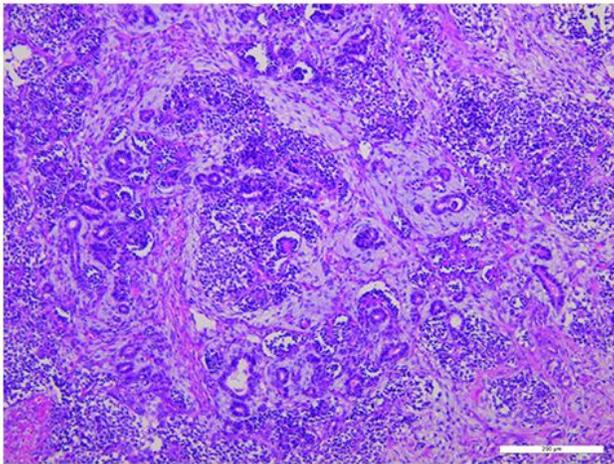
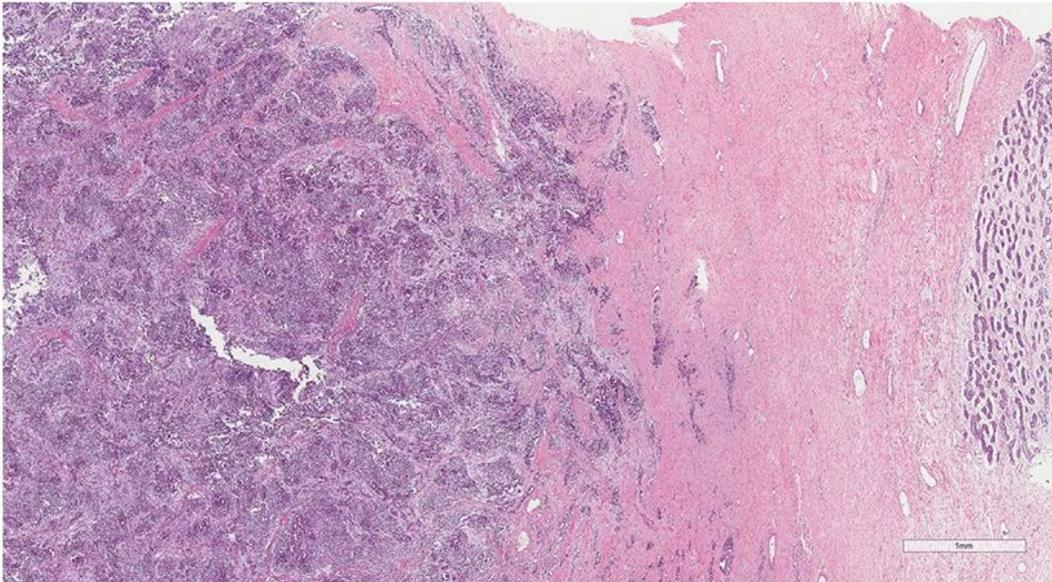
**Figure 1**

(A) Enhanced CT showed significant enlargement of the right testis, (B) Delayed enhancement with enhanced scanning, and enhanced angiography.



**Figure 2**

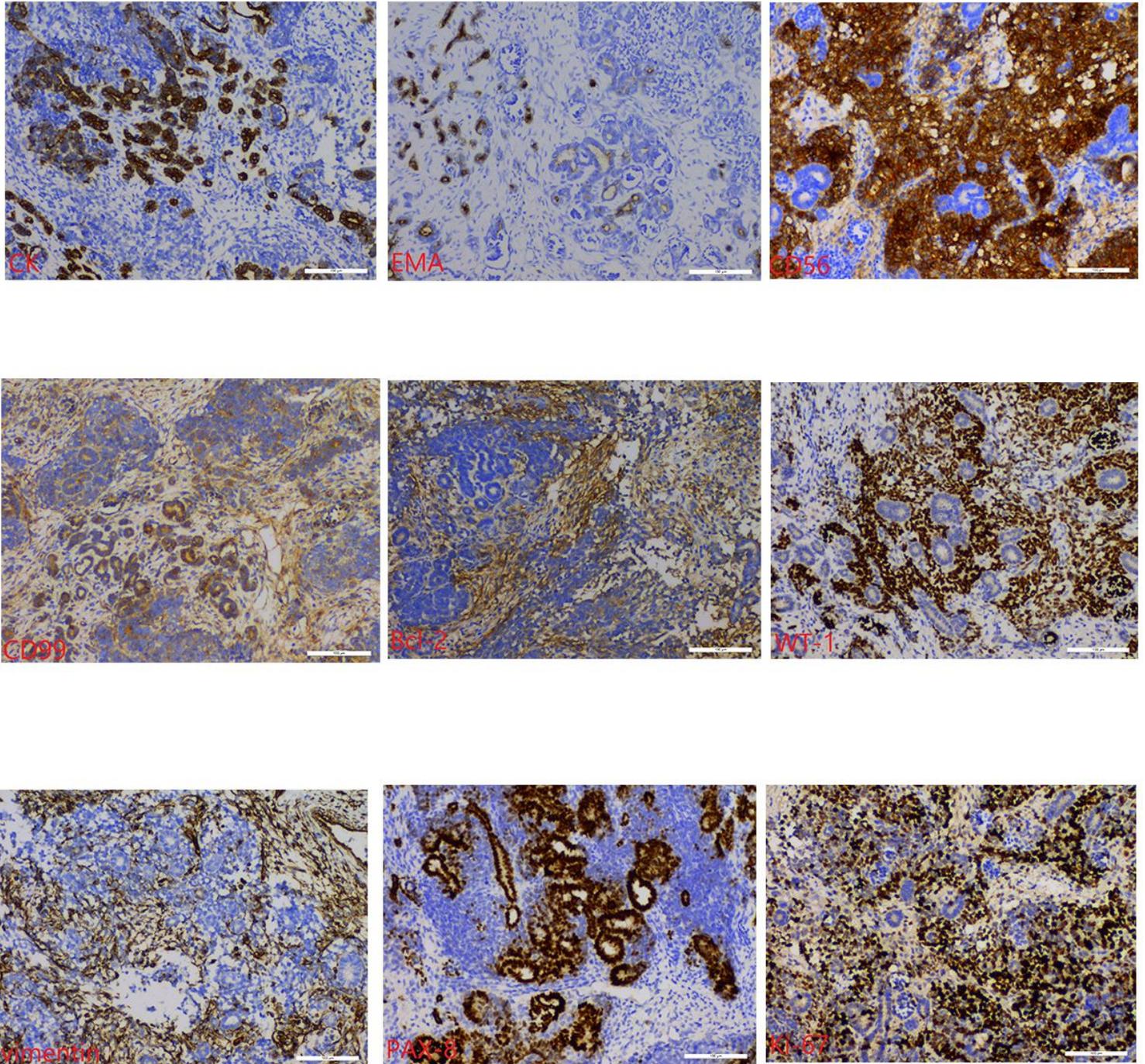
(A)The mass was a solid mass encapsulated by a fibrous capsule, (B)The cut section showed a milky white cut-surface.



**Figure 3**

(A) Microscopic examination showed the characteristics of the simultaneous exhibiting triphasic pattern of Wilms tumor. The testicular tissue was entirely supplanted by neoplastic elements with proof of immature testicular tissue in the peripheral area of the tumor (HE  $\times 200$ ). (C) Microscopic evaluation of the specimen revealed a triphasic pattern of Wilms tumor. (HE  $\times 100$ ), (D) Small blue cells were organized in serpiginous aggregates (blastemal component), strictly constrained by focal spindling and intervening

collagenous bundles separately from the surrounding stromal elements. There are also a few small tubules lined by primitive cuboidal cells and a small area of nephrogenic rest at the periphery. (HE ×200)



**Figure 4**

Immunohistochemically, the stromal component was positive for vimentin and SMA. The blastemal component was positive for vimentin positive for WT1, CD56, CD99, while the epithelial component was positive for EMA, CK, PAX-8, and vimentin. The Ki-67 index in the blastemal component is about 80% and

in the epithelial and mesenchymal approximately 10%. NSE, S100, Inhibin, LCA, RCC, SALL4, P53, myogenin, SALL4, CgA, Syn, S-100, and desmin were negative. (EnVision method, ×200).

## Supplementary Files

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