



## Rhabdomyosarcoma of the kidney: A case report

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### ABSTRACT

Rhabdomyosarcoma is a mesenchymal malignancy. It can occur anywhere in the body, but is extremely rare in the kidney. Here, we report a rare case of rhabdomyosarcoma of the kidney in a 19-year-old female who presented with right lumbar pain. Abdominal computed tomography (CT) suggested a huge retroperitoneal mass (approximately 18 × 14 cm). Preoperative neoadjuvant chemotherapy was performed through two preoperative cycles. After admission to our department for further evaluation, enhanced abdominal CT showed a mass of the right kidney (9.6 cm × 7.1 cm). The mass was successfully removed by surgery and the patient achieved a complete recovery.

### 1. Introduction

Rhabdomyosarcoma (RMS) is a highly malignant mesenchymal tumor that is rare in adults, accounting for 2%–5% of adult soft tissue tumors, and about 20% occur in the genito-urinary system.<sup>1</sup> At this site, RMS occurs most commonly in the bladder, prostate, testis and adjacent to the testis, but is rare in the kidney. Preoperative diagnosis of renal rhabdomyosarcoma is difficult, and based mainly on imaging, pathology and immunohistochemical markers. Surgery is the main treatment method, supplemented by radiotherapy and chemotherapy. Here, we describe a case of renal RMS admitted to our hospital to provide clinical guidance for this disease.

### 2. Case presentation

The 19-year-old female patient was admitted to the hospital on February 18, 2022. A right renal mass (approximately 2 × 3 cm) had been detected in physical examination 2+ years previously at a different hospital. No treatment was given at that time. In addition, the patient reported right lumbago without obvious causes and intermittent colic for 1+ years. Abdominal computed tomography (CT) suggested a huge retroperitoneal mass (approximately 18 × 14 cm). Radical surgery was not performed due to the huge mass and serious adhesion with surrounding tissues. After laparoscopic exploration, PET-CT, and pathological examination of a biopsy, pleomorphic rhabdomyosarcoma of the right kidney was diagnosed. Right renal artery embolization chemotherapy was performed, followed by two cycles of chemotherapy. Subsequent abdominal CT indicated a reduction in the tumor size. After

admission to our department for further evaluation, enhanced abdominal CT showed a mass of the right kidney (9.6 cm × 7.1 cm) consistent with pleomorphic rhabdomyosarcoma (Fig. 1). On February 25, radical resection of the right kidney tumor was performed by laparoscopy. No obvious normal kidney tissue was found during the operation, and the tumor (14 cm × 11.5 cm × 6.5 cm) showed serious adhesion to the surrounding tissues (Fig. 2). Post-surgery, the patient's condition was stable and a complete recovery was achieved. Postoperative pathological results indicated right renal malignancy accompanied by necrosis based on HE staining and immunohistochemical analysis of the following mesenchymal and myogenic markers: Vimentin (partial+), Ki-67 (70%–90%+), SMA (weak+), desmin (partial+), MyoD1 (partial+), Bcl-2 (90%+), CD56 (partial+), CD4 (partial+), INI-1 (+) (Fig. 3).

### 3. Discussion

RMS, which is a primitive mesenchymal malignant tumor with skeletal muscle differentiation tendency or derived from skeletal muscle, is commonly seen in patients aged under 15 years, accounting for 5%–10% of solid tumors in children.<sup>2</sup> In contrast, RMS is rare in adults, with the genito-urinary system as the second most common site<sup>1</sup> and a higher incidence in males than females.<sup>3</sup> Primary RMS is a very rare tumor, accounting for 1%–3% of primary renal malignancies,<sup>2</sup> and this disease rarely occurs in adults. Reports of the disease are rare, and most describe individual cases. Although there are no distinct clinical manifestations, renal rhabdomyosarcoma is manifested mainly as an abdominal mass with symptoms such as abdominal pain and signs such as gross hematuria, which become obvious due to its strong invasion and rapid

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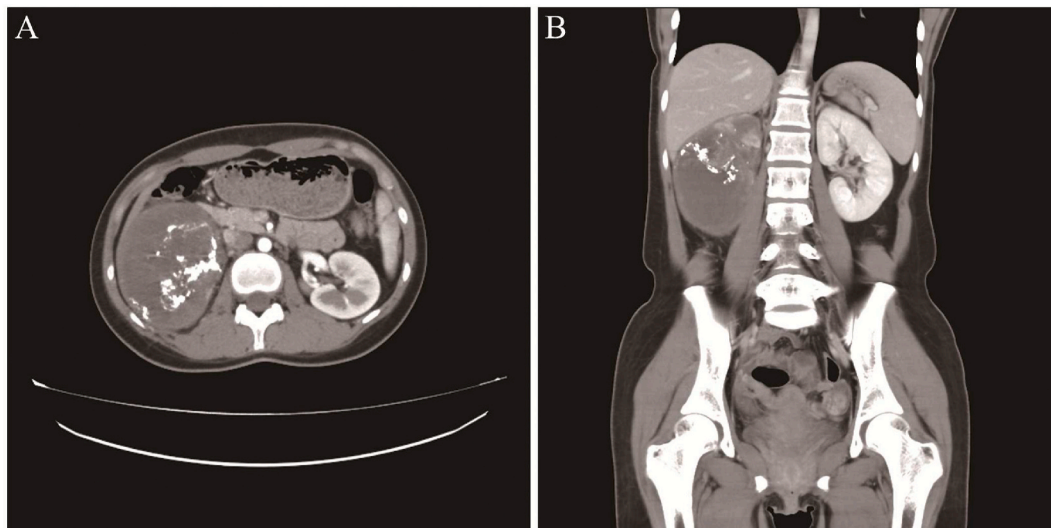


Fig. 1. Computed tomography shows right renal mass.

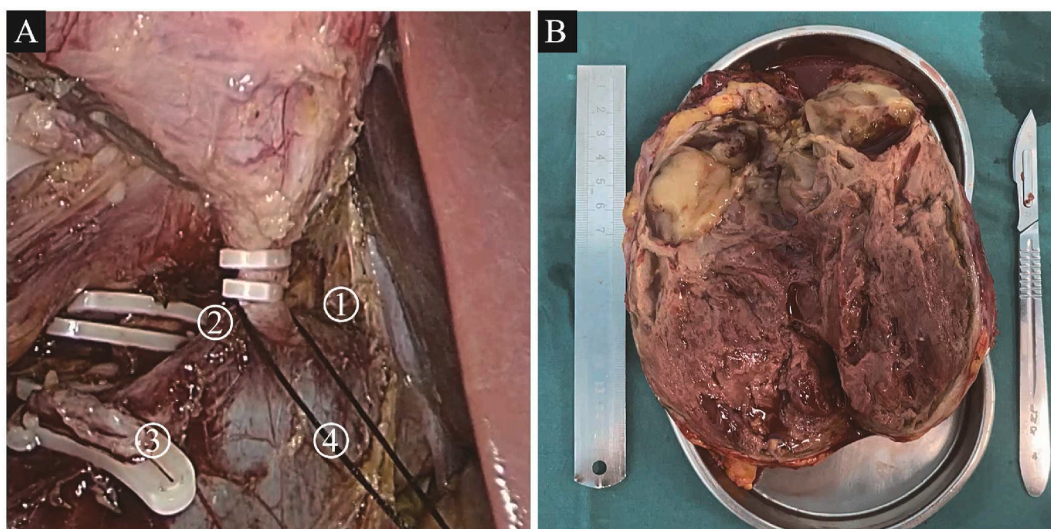


Fig. 2. Intraoperative conditions, kidney and tumor tissue

Fig. 2A: ①right renal vein ②right renal artery ③Genital vein ④postcava; Fig. 2b: shows the kidney tumor after separation and resection, which is about 14cm × 11.5cm × 6.5cm in size. The tumor area is soft and grayish white like fish meat, and some areas are grayish brown like necrosis. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

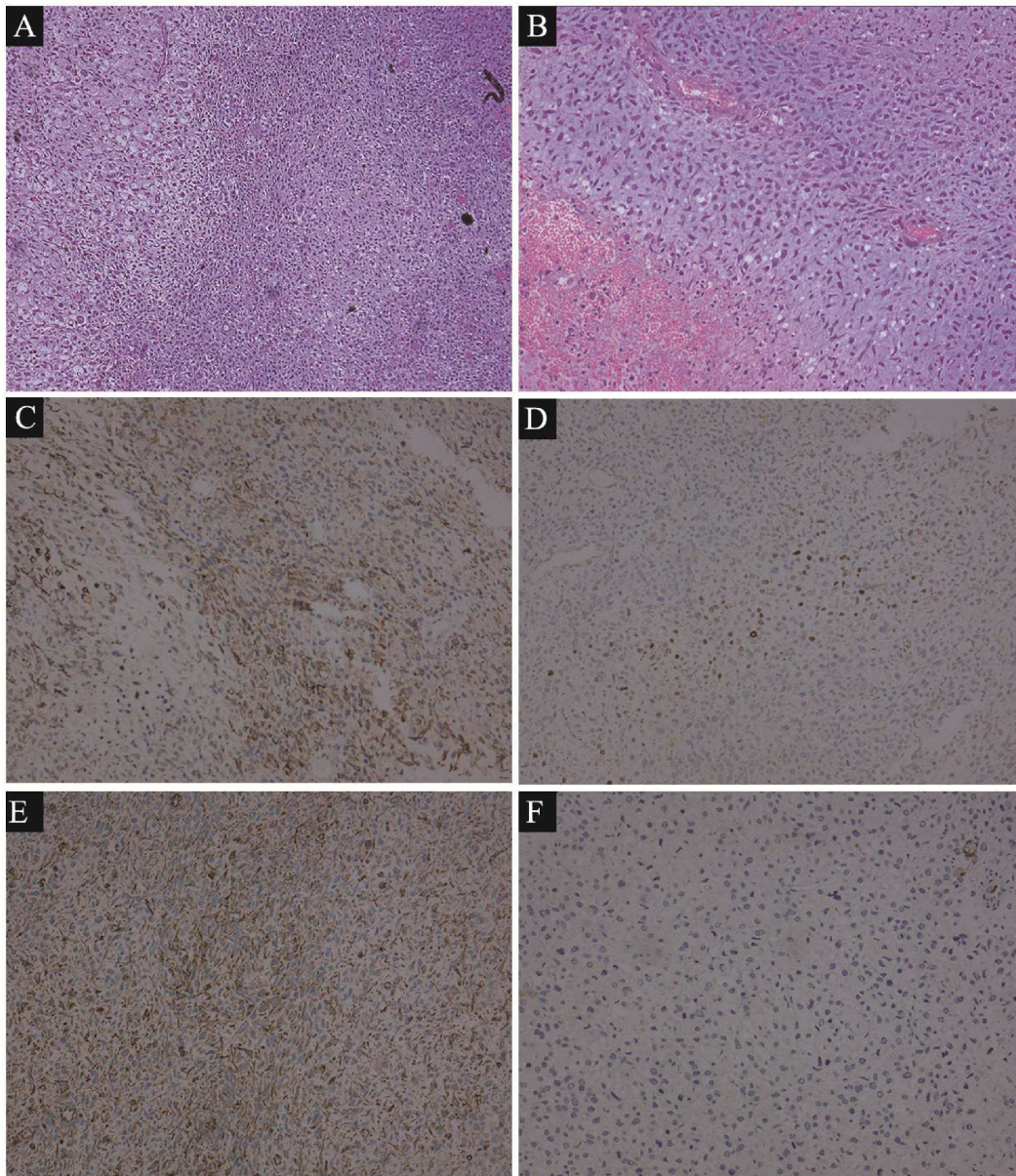
growth. In our case, the main clinical manifestations were right lumbago and an abdominal mass, without gross hematuria or other symptoms.

The diagnosis of renal RMS is based mainly on imaging, pathology and immunohistochemical markers. In this case, the patient was diagnosed with a malignant right renal tumor by PET-CT performed at another hospital. Our tumor biopsy analysis revealed a recurrence of the polymorphic RMS (PRMS) of the right kidney. This was supported by detection of a large mass of the right kidney in enhanced CT imaging of the whole abdomen. In general, the kidney and the tumor tissue were not clearly demarcated. The tumor was large and consisted of gray tissue with areas of texture and necrosis in some areas. HE staining showed diffuse distribution and obvious atypia of tumor cells, with partial necrosis of the tumor tissue. Immunohistochemical analysis showed that the tumor was positive for mesenchymal and myogenic markers. Based on relevant these results, the renal tumor was diagnosed as RMS.

The treatment of renal RMS is multi-modal, based mainly on a comprehensive plan consisting of surgery, supplemented by radiotherapy and chemotherapy. However, complete surgical resection of the

tumor is the ultimate goal to improve the survival rate of patients. For patients with incomplete resection or a positive resection margin, radiotherapy can reduce the risk of local recurrence. However, since many patients with RMS of the genito-urinary system are relatively young, care should be taken to avoid genito-urinary dysfunction caused by radiotherapy. Targeted radiotherapy may reduce radiation-related complications, such as external proton radiotherapy or brachytherapy.<sup>4</sup> Systemic chemotherapy is the basis of multimodal treatment for RMS patient,<sup>5</sup> with vincristine, actinomycin, and alkylating agents (ifosfamide or cyclophosphamide) as the main drugs. For very high-risk RMS patients, such as those in disease remission, maintenance chemotherapy is also recommended after the end of consolidation chemotherapy. Compared with bladder and prostate cancers, the survival rate of adult renal RMS is worse. The age of onset and degree of disease are related to the prognosis, and surgical intervention and chemotherapy contribute to improved survival rates.<sup>1</sup> Therefore, early diagnosis and intervention are essential to improve the survival rate of this rare malignancy in adulthood.





**Fig. 3.** Pathological section of renal rhabdomyosarcoma ( HE  $\times 100$  ) and relevant immunohistochemical markers (  $\times 100$  ).

Note : It shows the tumor cells were diffuse and heteromorphic(A), It shows the necrosis of tumor tissue(B).Immunohistochemical staining shows cytoplasmic positivity for Des(C) , MyoD1(D),Vimentin(E); S-100(F).

In this case, the patient received preoperative chemotherapy after definite diagnosis followed by radical resection of the right kidney tumor by laparoscopy. After review of the radical resection treatment, the patient recovered well and the pathology report did not suggest lymph node metastasis. Postoperative adjuvant chemotherapy was administered, but late recovery and survival require further follow-up.

#### 4. Conclusion

Adult renal RMS has a high degree of malignancy, strong infiltration and poor prognosis. Early detection, diagnosis and treatment can achieve a better curative effect. The rarity of adult renal RMS has hindered the study of this disease and blocked progress in the diagnosis and treatment of this tumor. Further research focused on molecular-targeted therapeutic strategies is expected to facilitate the development of more

effective chemotherapy drugs with fewer side-effects to improve the survival rate of patients.

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