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Primary embryonal rhabdomyosarcoma of the kidney in an adult: A case report

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► *Case report*

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ABSTRACT

Primary rhabdomyosarcoma (RMS) of the kidney is a rare malignant mesenchymal tumor with an aggressive clinical course. Adult renal RMS is typically a pleomorphic histologic subtype and only a few cases have ever been reported. We herein present a new case of renal RMS of the embryonal histologic subtype in a 26-year-old woman.

Keywords: *Embryonal rhabdomyosarcoma, kidney, tumor, adult.*

INTRODUCTION

Rhabdomyosarcoma (RMS) is a malignant skeletal muscle neoplasm. RMS can occur anywhere in the body and is the most common childhood sarcoma [1]. Pediatric RMS is an aggressive neoplasm with deregulated cell cycle checkpoints, high genome instability, and oncongene amplification such as MYC family members [2]. However, RMS rarely occurs in adults. Moreover, sarcomas of the kidney are rare soft tissue tumors comprising approximately 1% of all primary renal malignancies in adults [3]. Primary renal RMS of adults is characterized by pleomorphic histology and poor prognosis, but is extremely unusual [4]. Here, we report a case of adult primary renal RMS of the embryonal histopathologic subtype.

Case report

A 26-year-old woman was admitted to the Affiliated Hospital of Qingdao University Medical College in Qingdao, China, with complaints of pain in her right flank and the presence of gross hematuria for two days. She also complained of nausea and vomiting. The physical examination was unremarkable except for mild tenderness in right flank. Head and neck, chest, and cardiovascular examinations were all normal. Urinalysis revealed 25-30 RBCs/HPF and mild proteinuria (1+). Other laboratory values were within normal ranges.

An abdominal ultrasound demonstrated a solid hetero-echoic mass of 6.4×4.4 cm in the upper pole of the right kidney (figure 1). The mass displayed an irregular outer contour that protruded slightly from the renal parenchyma with foci of cystic necrosis. The tumor appeared to invade into the renal hilum. Hetero-echoic nodules could be vaguely seen in the renal hilum area, but no obvious invasion of the renal vein and inferior vena cava was found. Color Doppler flow imaging (CDFI) showed sparse blood flow within the tumor (figure 2). The diagnosis of a right renal tumor with upper sinus invasion was made.

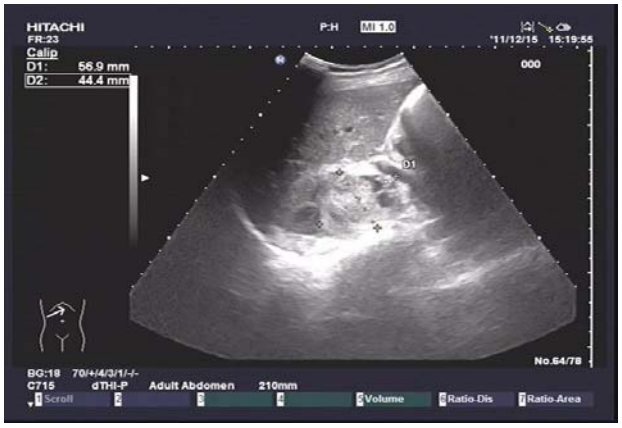

Figure 1. Ultrasound demonstrated a solid hetero-echoic mass in the upper pole of the right kidney. The mass had an irregular outer contour and foci of cystic necrosis. The tumor appeared to invade into the renal hilum.

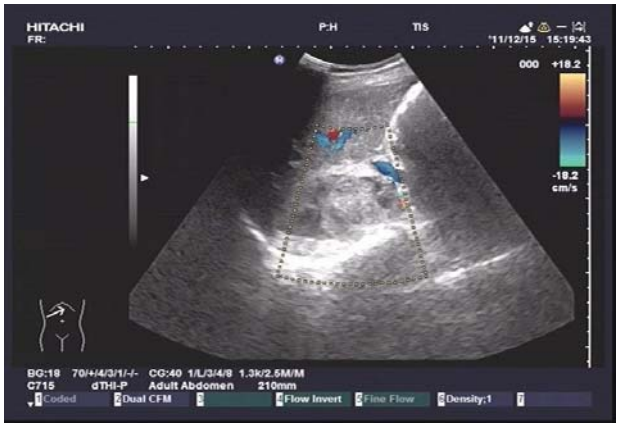

Figure 2. CDFI showed sparse blood flow within the tumor and no flow signal was found in the area of cystic necrosis.

MATERIALS AND METHODS

Computerized tomography (CT) showed a solid mass of 5.2×4.3cm protruding from the upper pole of the right renal parenchyma, which demonstrated heterogeneous delayed-enhanced mode. The right renal pelvis and upper ureter showed some expansion, but no swelling of the retroperitoneal lymph nodes was found. Preoperative diagnosis of a right renal tumor was subsequently made. A scan of the liver and spleen showed no clear invasion of the liver by the tumor and no metastatic deposits. A chest radiograph and CT scan did not show lung metastases, and a bone scan was normal.

A right radical kidney resection was performed. At the time of surgery, the tumor originating from the upper pole of the right kidney was found to be a gray mass with a smooth surface and pseudocapsule. The tumor tissues appeared to be fish-like and mixed with necrotic areas. Although the tumor had invaded the renal hilum and local vessels, the renal capsule and ureter were free of tumor.

Gross pathologic examination showed a 4cm gray solid mass with cystic and necrotic areas involving the upper renal pole. Light microscopy revealed a malignant tumor composed of large haphazardly arranged cells of various shapes with abundant and deeply eosinophilic cytoplasm. Cytoplasm cross-striations were identified in the spindle-shaped and tadpole-shaped cells (figure 3). There were numerous mitotic figures and areas of necrosis. Immunohistochemistry showed positive staining for myogenin (figure 4), smooth muscle actin (SMA), and desmin, but negative staining for cytokeratin, EMA, CD10, S-100 protein, and vimentin. These results indicated that the tumor

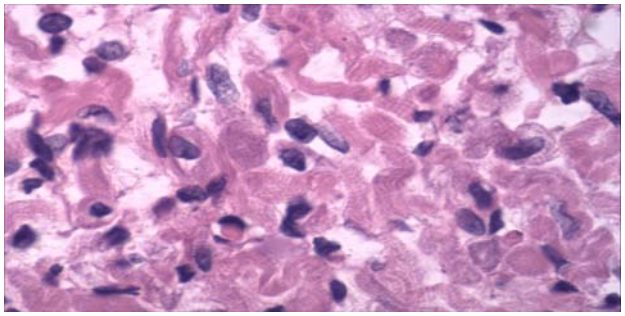

Figure 3. The spindle-shaped and tadpole-shaped tumor cells with abundant and deeply eosinophilic cytoplasm. Cytoplasmic cross-striations were identified in tumor cells. There were numerous mitotic figures (×400).

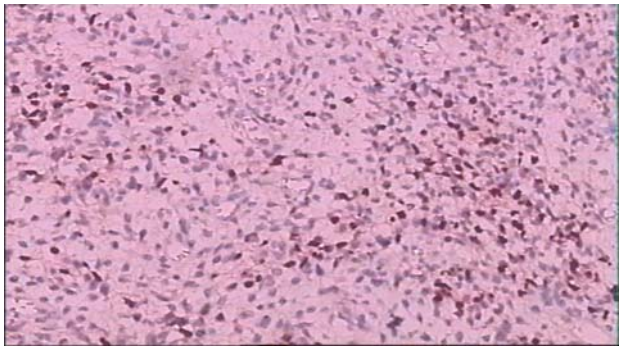

Figure 4. Representative immunohistochemical micrographs showing positive staining for myogenin in the tumor cells (×100).

originated from skeletal muscle, and the final diagnosis of embryonal RMS of the right kidney was made.

Postoperatively, the patient showed an unremarkable recovery and was scheduled to begin combination chemotherapy with Vincristine, Actinomycin D, and Cyclophosphamide with adjunctive 3-dimensional conformal radiotherapy as an outpatient. Four months after her original diagnosis, the patient was alive with no evidence of metastatic disease.

DISCUSSION

Rhabdomyosarcoma (RMS) is an aggressively malignant soft tissue tumor and is thought to derive from mesenchymal cells already committed to becoming skeletal muscle cells [5]. The majority of RMS tumors express high levels of early inducers of muscle differentiation, such as MyoD and myogenin[6]. RMS can be classified into different subtypes based on histologic features, where the main subtypes are: alveolar RMS (ARMS), embryonal RMS (ERMS), and pleomorphic RMS [7]. The embryonal and alveolar subtypes are frequently found in the pediatric age group, whereas the pleomorphic subtype is most often seen in adults [8-10]. RMS accounts for 5-8% of childhood tumors, making it the most common soft tissue tumor in the pediatric population [1]. It can arise in any location, but 60% of pediatric cases are found in the head and neck and urogenital tract, while very few are found in skeletal muscle [8]. RMS in adults arises mainly in the large skeletal muscles, whereas the lower extremity is the most frequent site, followed by the trunk [9].

Renal RMS in adults is extremely uncommon. To date and to the best of our knowledge, only 11 cases have been reported in the English language literature [9,10]. Most of these adult RMS cases were of the pleomorphic histologic subtype. The most comprehensive report to date is a case report and review of 7 cases by Grignon *et al.* [11]. The most recent case report is a 43-year-old woman with pleomorphic RMS of the left kidney [4]. Due to the lack of a standardized and effective treatment, most of these reported patients died shortly following diagnosis.

In our case, abdominal sonography and CT with contrast showed the location, size, border, texture, and invasive properties of the tumor. For treatment options, further characteristic imaging is needed to confirm our diagnosis. Light microscopic examination and immunochemical staining supported the final diagnosis of primary embryonal renal RMS. Furthermore, our case supported the criteria for the diagnosis of primary renal sarcoma proposed by Grignon *et al.* [11]. First, our patient did not have a previous history of sarcoma elsewhere, eliminating the possibility of metastatic disease. Second, imaging examinations and histopathologic findings demonstrated that the tumor arose in renal parenchyma rather than invasion from outside the kidney. Finally, microscopic examination and immunopathology excluded a sarcomatoid renal cell carcinoma.

Histologically, the primary renal RMS is a high-grade tumor that needs to be differentiated from other malignant lesions, such as sarcomatoid renal cell carcinoma, metastatic carcinoma or melanoma, and rhabdoid tumor. Immunohistochemistry and clinical features of the tumor may help to narrow the list of differential diagnoses.

In summary, we presented a rare case of primary embryonal RMS of the right kidney in an adult woman based on the histopathology and immunohistochemistry. Due to so few reports of renal RMS, the experience in treating and managing adult patients is limited. Radical nephrectomy remains an effective therapeutic option for localized renal disease, and adjuvant radiotherapy might reduce the risk of local recurrence. The prognosis of adult renal RMS was generally very poor. To improve the evaluation and management of future cases, further studies are needed to elucidate its histogenesis.

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