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CASE REPORT

Extrarenal malignant rhabdoid tumors: radiologic findings with histopathologic correlation

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Abstract

Extrarenal malignant rhabdoid tumors (MRT) are extremely rare. There have been only a few cases of MRT described in the literature and even fewer in the radiology literature. We present rare uterine and hepatic MRT and their computed tomography, magnetic resonance, and ultrasound imaging features with histopathologic correlation. Although the final diagnosis is based largely on histopathology, radiologists should include MRT in their differential considerations, regardless of tumor location, when they are dealing with an extremely aggressive tumor of early childhood.

Keywords: *Malignant rhabdoid tumor; CT; MR; uterine rhabdoid tumor.*

Introduction

Rhabdoid tumor was first described in the kidneys and was initially thought to be a sarcomatous variant of Wilms tumor[1]. Renal rhabdoid tumor has been described as a solid tumor that is seen usually only in childhood, with most cases occurring in the 1st year of life[2]. These tumors are usually large at presentation and show necrosis and calcification more commonly than Wilms tumor. Affected patients have a dismal 18-month survival rate of only 20%[3]. Interesting features of renal rhabdoid tumors include their association with hypercalcemia, the presence of lobulation at radiology, and occasional subcapsular hematoma formation[1,3].

The two largest series of extrarenal soft-tissue tumors with a rhabdoid phenotype in the pathology literature were reported by Parham *et al.*[4] and Kodet *et al.*[5] Extrarenal rhabdoid tumors have been described in the liver, brain, tongue, neck, chest, heart, pelvis, extremities, and several other sites[5]. These tumors typically occur in infancy; the median patient age in one large series was 20 months. However, patient age ranges from 3 weeks to 50 years[4]. The tumors have a 1.5:1 male predilection[4], and death occurs an average of 6 months after diagnosis. Data concerning the 5-year survival rate are not available.

The imaging characteristics of soft-tissue rhabdoid tumors have yet to be determined, largely because of the rarity of these tumors[6,7]. Recently, Garcés-Iñigo *et al.*[8] described the largest case series in the radiology literature; no specific imaging features were observed that could prove diagnostic for these tumors, however, Garcés-Iñigo *et al.*[8] did show that these tumors have a tendency to be large and hypodense on CT, and show a heterogeneous hyperintensity on T2-weighted (W) magnetic resonance (MR) imaging. To our knowledge, only a few cases of hepatic and none of malignant uterine rhabdoid tumors have been described in the radiology literature previously.

Case report 1

The patient was a 19-month-old white female with no relevant past medical history who presented with dysuria. Ultrasound imaging revealed bilateral hydronephrosis possibly as a result of bladder outlet obstruction from a

large heterogeneously hyperechoic mass arising from the uterus; color Doppler imaging showed tumor vascularity (Fig. 1).

The patient subsequently underwent MR imaging of the pelvis, which showed a large mass arising in the uterus that was heterogeneous and iso- to hyperintense on T2W imaging and isointensity on T1W sequence (Fig. 2). Post-contrast T1W MR imaging showed an avid heterogeneous enhancement pattern with areas of low intensity suggestive of necrosis. The uterus appeared enlarged with diffusely irregular wall displacing the bladder anteriorly secondary to this large irregular mass (Fig. 3).

The patient underwent a biopsy which revealed a rare uterine rhabdoid tumor.

Subsequent CT imaging of chest, abdomen, and pelvis was performed to evaluate for metastatic disease; multiple hypodense rounded lesions distributed throughout the liver were consistent with metastasis (Fig. 5). A whole body nuclear medicine bone scan (not shown) was also performed; this showed a focally prominent activity overlying the sacrum that was felt to be either a result of small dystrophic calcification within the mass or extension of the mass into the anterior sacral cortex.

Follow-up MR imaging revealed multiple hyperintense lesions in the liver on T2W sequence which appeared hypointense on T1W sequence. Post-contrast images revealed moderate heterogeneous enhancement (Fig. 4). The findings were suggestive of metastatic disease.

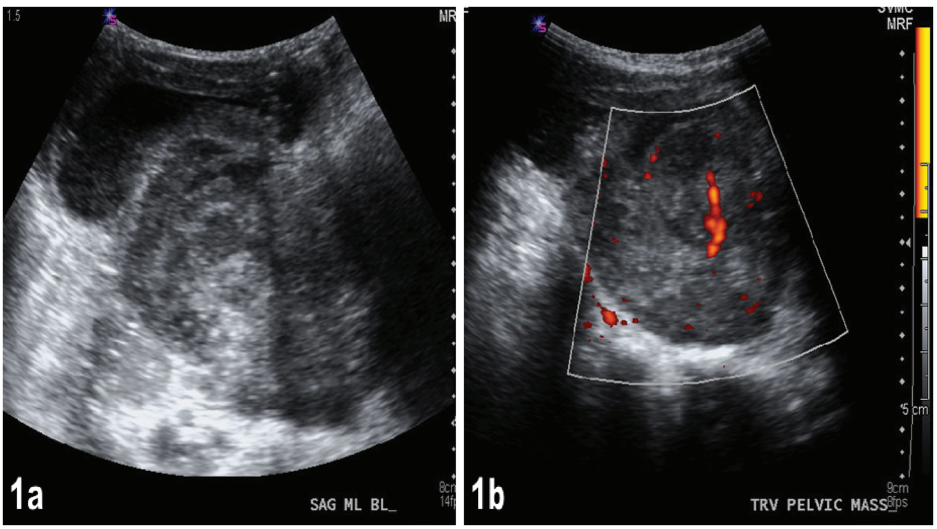

Figure 1 **Grayscale and color Doppler images of the pelvis showing a large heterogeneous echotexture solid uterine mass in case 1 with internal vascularity and mass effect on the bladder which appears anteriorly displaced.**

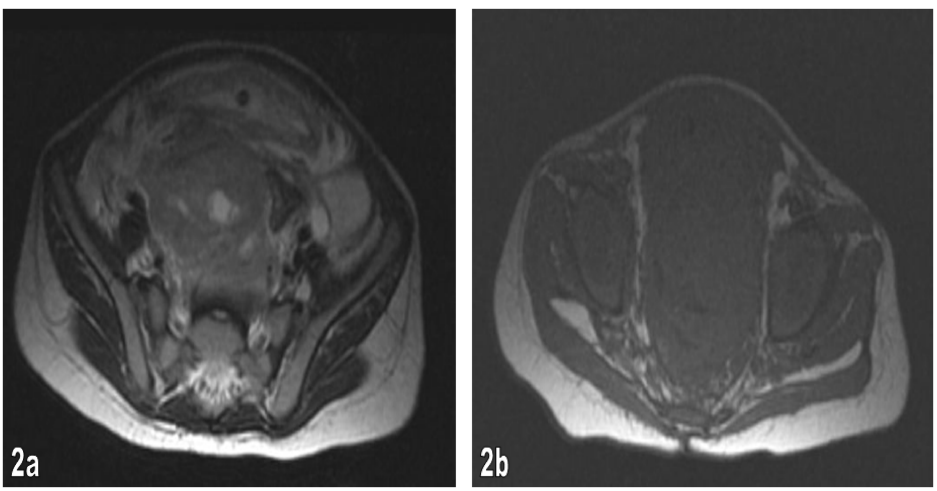

Figure 2 **Axial T1W MR image (b) shows a large isointense uterine mass in case 1 with mass effect and anterior displacement of the bladder. On axial T2W MR image (a), the uterine mass appears heterogeneous with areas of hyperintensity.**

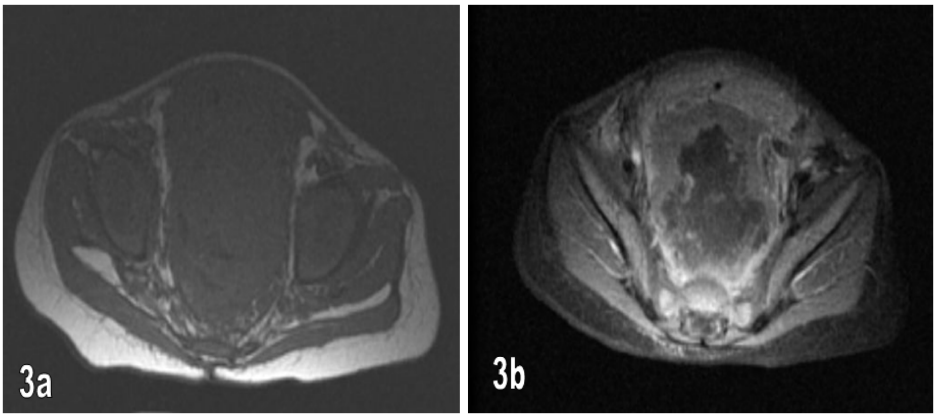

Figure 3 **Pre- and post-contrast T1W MR images of the uterine mass in case 1. Post-contrast axial T1W MR image (b) showing heterogeneously avid enhancement with areas of hypointensity suggestive of necrosis.**

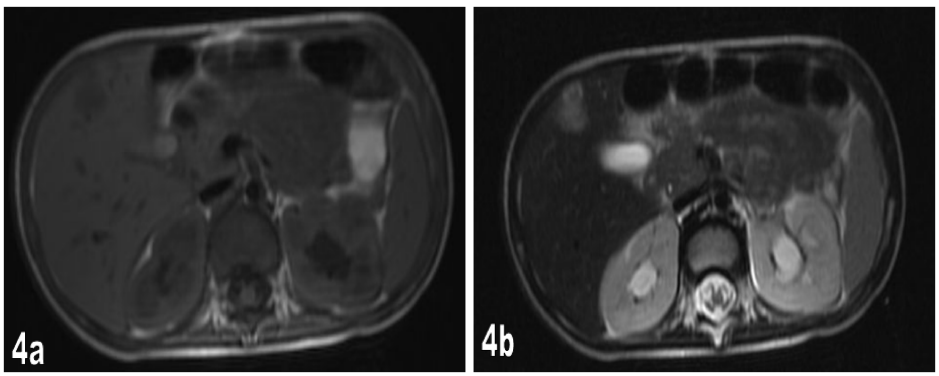

Figure 4 **An isointense mass on axial T1W MR imaging (a) in the anterior segment of the right lobe of the liver in case 1, which appears heterogeneously hyperintense on axial T2W MR imaging (b) suggestive of metastatic disease.**

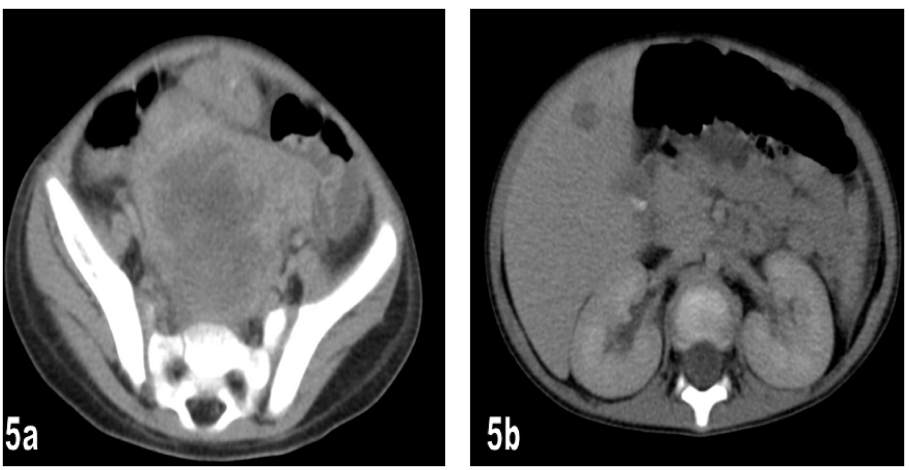

Figure 5 **Axial CT post-contrast images show a large heterogeneous, predominantly hypodense uterine mass (a) in case 1 with mass effect and displacement of adjacent structures. A hypodense lesion is seen in the right lobe of the liver most consistent with metastatic disease (b).**

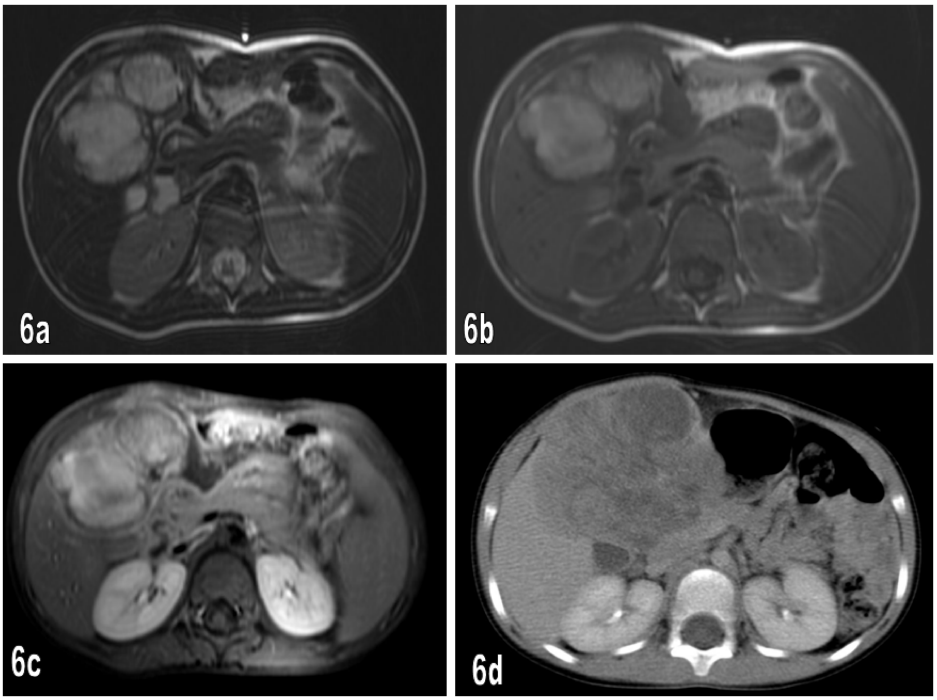

Figure 6 **Axial T2W MR image (a) shows a multilobulated heterogeneously hyperintense mass in case 2, which appears hyperintense on T1W MR image (b) involving the right lobe of the liver. Axial T1W MR post-contrast image shows heterogeneously avid enhancement of this mass, which appears lobulated (c). Axial CT post-contrast image shows a large macrolobulated heterogeneous and predominantly hypodense mass involving the right lobe of the liver in case 2 with areas of low density suggestive of necrosis.**

Case report 2

The patient was a healthy 3-year-old white male with no significant past medical history who presented with right upper quadrant pain, intermittent fever, and distended abdomen. Physical examination revealed tenderness to palpation in the right upper quadrant and hepatomegaly. Initial imaging with CT was performed and revealed a well-circumscribed, large, heterogeneous, hypodense mass with areas of necrosis within the anterior segment of the right lobe of the liver (Fig. 6d).

MR imaging showed a large heterogeneous, hyperintense, mutilobulated liver mass involving the inferior aspect of the right lobe of the liver on T2W imaging with mass effect on the main portal vein (Fig. 6a). Post-contrast T1W imaging showed heterogeneous contrast enhancement (Fig. 6c).

Serum levels of ferritin, alpha-fetoprotein, and beta-human chorionic gonadotropin were within limits of normal, however, the patient did have increased lactate dehydrogenase levels of approximately 1371 IU/l. Further CT imaging of the chest and brain did not reveal any metastatic disease. The tumor was judged to be surgically unresectable because of its close proximity to porta hepatis. An open wedge biopsy was performed and tumor histology confirmed a rare rhabdoid tumor of the liver.

Discussion

Malignant rhabdoid tumors (MRTs) are highly aggressive neoplasms that most commonly occur in the kidney of young children. A few cases of primary MRT occurring in extrarenal sites have been reported, particularly in the soft tissues. MRT was first described by Beckwith and Palmer in 1978 as a rhabdomyosarcomatoid variant of Wilms tumor with a poor prognosis[1].

Most MRTs occur during the first year of life[9]. The median age at presentation is about 11 months and is distributed over early infancy and later childhood and tails off into adult life[10,11]. The course of extrarenal MRT is short and its prognosis is very poor. Less than 50% of patients with this neoplasm have survived for 5 years without tumor recurrence, regardless of the therapy used[12–14].

Our first case was a malignant and highly aggressive tumor of the endometrium, which corresponded to Beckwith's restrictive morphological criteria for MRT[14], with all the main histological, immunohistochemical and ultrastructural characteristics of the entity.

Radiologically, MRTs are large heterogeneous tumors on CT and MRI at the time of presentation[1,8,15]. They demonstrate contrast enhancement and areas of necrosis. They can be locally invasive. Extrarenal MRTs are highly aggressive neoplasms and patients may have metastatic

disease at the time of presentation. The liver may be the primary site of metastatic involvement. Hypodensity on CT seems to be a consistent feature of these aggressive tumors, as seen in our cases and in a recent case series published by Garcés-Iñigo *et al.*[8]. Our cases also showed heterogeneous hyperintensity on T2W MR and heterogeneous enhancement as seen in the case series by Garcés-Iñigo *et al.*[8].

In summary, although the diagnosis is largely based on histopathology, radiologists should include MRT in their differential considerations, regardless of tumor location, when they are dealing with an extremely aggressive tumor of early childhood.

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References

[1] Sisler C, Siegel M. Malignant rhabdoid tumor of the kidney: radiologic features. Radiology 1989; 172: 211–2.

[2] Parham D. Pediatric neoplasia: morphology and biology. Philadelphia, PA: Lippincott Raven; 1996, p. 54–5.

[3] Chung C, Lorenzo R, Rayder S, *et al.* Rhabdoid tumors of the kidney in children. AJR 1995; 164: 697–700.

[4] Parham D, Weeks D, Beckwith JB. The clinicopathologic spectrum of putative extrarenal rhabdoid tumors. Am J Surg Pathol 1994; 18: 1010–29.

[5] Kodet R, Newton W, Sachs N, *et al.* Rhabdoid tumors of soft tissues. Hum Pathol 1991; 22: 674–84. doi:10.1016/0046-8177(91)90289-2.

[6] Howlett DC, King AP, Jarosz JM, *et al.* Imaging and pathological features of primary malignant rhabdoid tumours of the brain and spine. Neuroradiology 1997; 39: 719–23. doi:10.1007/s002340050494. PMid:9351109.

[7] Winger DI, Buyuk A, Bohrer S, *et al.* Radiology-Pathology Conference: rhabdoid tumor of the kidney. Clin Imaging 2006; 30: 132–6. doi:10.1016/j.clinimag.2005.09.027. PMid:16500545.

[8] Garcés-Iñigo EF, Leung R, Sebire JN, McHugh K. Extrarenal rhabdoid tumours outside the central nervous system in infancy. Pediatr Radiol 2009; 39: 817–22.

[9] Orbach D, Rey A, Oberlin O, *et al.* Soft tissue sarcoma or malignant mesenchymal tumors in the first year of life: experience of the International Society of Pediatric Oncology (SIOP) Malignant Mesenchymal Tumor Committee. J Clin Oncol 2005; 23: 4363–71. doi:10.1200/JCO.2005.12.009. PMid:15994146.

[10] Fanburg-Smith JC, Hengge M, Hengge UR, *et al.* Extrarenal rhabdoid tumors of soft tissue: a clinicopathologic and immunohistochemical study of 18 cases. Ann Diagn Pathol 1998; 6: 351–62.

[11] Brennan DM, Foot MBA, Stiller C. Where to next with extracranial rhabdoid tumours in children. Eur J Cancer 2004; 40: 624–6. doi:10.1016/j.ejca.2003.11.014. PMid:14962734.

[12] Perlman E, Ali S, Robinson R, *et al.* Infantile extrarenal rhabdoid tumor. Pediatr Dev Pathol 1998; 1: 149–52. doi:10.1007/s100249900019. PMid:9507040.

[13] Oda Y, Tsuneyoshi M. Extrarenal rhabdoid tumors of soft tissue: clinicopathological and molecular genetic review and distinction from other soft-tissue sarcomas with rhabdoid features. Pathol Int 2006; 56: 287–95. doi:10.1111/j.1440-1827.2006.01962.x. PMid:16704491.

[14] Hunt SJ, Anderson WD. Malignant rhabdoid tumor of the liver. A distinct clinicopathologic entity. Am J Clin Pathol 1990; 94: 645–8.

[15] Figarola M, Khader MS. Pediatric case of the day. Radiographics 1999; 19: 1693–5.