

CASE REPORT

Magnetic resonance imaging features of renal synovial sarcoma: a case report

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Date accepted for publication 21 January 2008

Abstract

Primary renal synovial sarcoma (SS) was first described in 2000 by Argani, with only a few subsequent cases being reported in the English literature. Herein, we describe a case of a 52-year-old woman who presented with right flank pain. Magnetic resonance imaging revealed a 6-cm mass in the lower pole of the right kidney. T1 and T2 weighted imaging revealed a heterogeneous mass with triple sign. There was post-contrast enhancement. Imaging, histology and immunostaining together made the diagnosis of SS of the kidney.

Keywords: Synovial sarcoma; kidney; renal; MRI.

Introduction

Synovial sarcoma (SS) most commonly affects the para-articular regions of the extremities of young adults^[1,2]. Occasionally, however, SS has been identified in less common locations, including the head and neck, heart, lungs and prostate^[3]. The first reported case of renal SS was described in 2000^[4].

Herein, we describe a case of primary renal SS, an exceedingly rare diagnosis, in a 52-year-old woman. The diagnosis was established by histology and immunostaining. To our knowledge, based on a review of the English literature, this marks the first case report describing the magnetic resonance (MR) features of synovial sarcoma of the kidney.

Case report

A 52-year-old female presenting with a history of right flank pain underwent computed tomography (CT) examination which revealed an amorphous partially cystic right lower pole renal mass with heterogeneous enhancement. Further workup with magnetic resonance imaging revealed a 6.0 × 5.9 × 6.1 cm mass in the lower pole of

the right kidney. Three-plane localizer, coronal single shot fast spin echo (SSFSE) T2, axial in and out of phase, axial fat suppressed T2 weighted, axial 5-min and 10-min delayed, and axial 3D liver acquisition with volume acceleration (LAVA) sequences were acquired. On T1 weighted imaging (Fig. 1A), the mass was heterogeneous, with the posterior region having similar intensity to skeletal muscle and the anterior region with higher signal intensity. On chemical shift imaging, the tumor showed no areas of signal intensity loss. Axial T2 weighted imaging revealed marked heterogeneity with areas of high, intermediate, and low signal, known as the 'triple sign'^[1] (Fig. 1B). The mass contained multiple areas of high T2 signal emanating from the center of the mass, with surrounding intermediate and low T2 signal intensity regions (Fig. 1E). These low T2 areas showed enhancement on post-contrast images (Fig. 1C,D). The posterior aspect of the mass appeared solid. Our prospective diagnosis based on these MR imaging findings was renal cell carcinoma.

The patient underwent right nephrectomy with a pathological diagnosis of high-grade, poorly differentiated spindle-cell synovial sarcoma. Immunohistochemical staining showed focal positivity for cytokeratin CAM 5.2 and

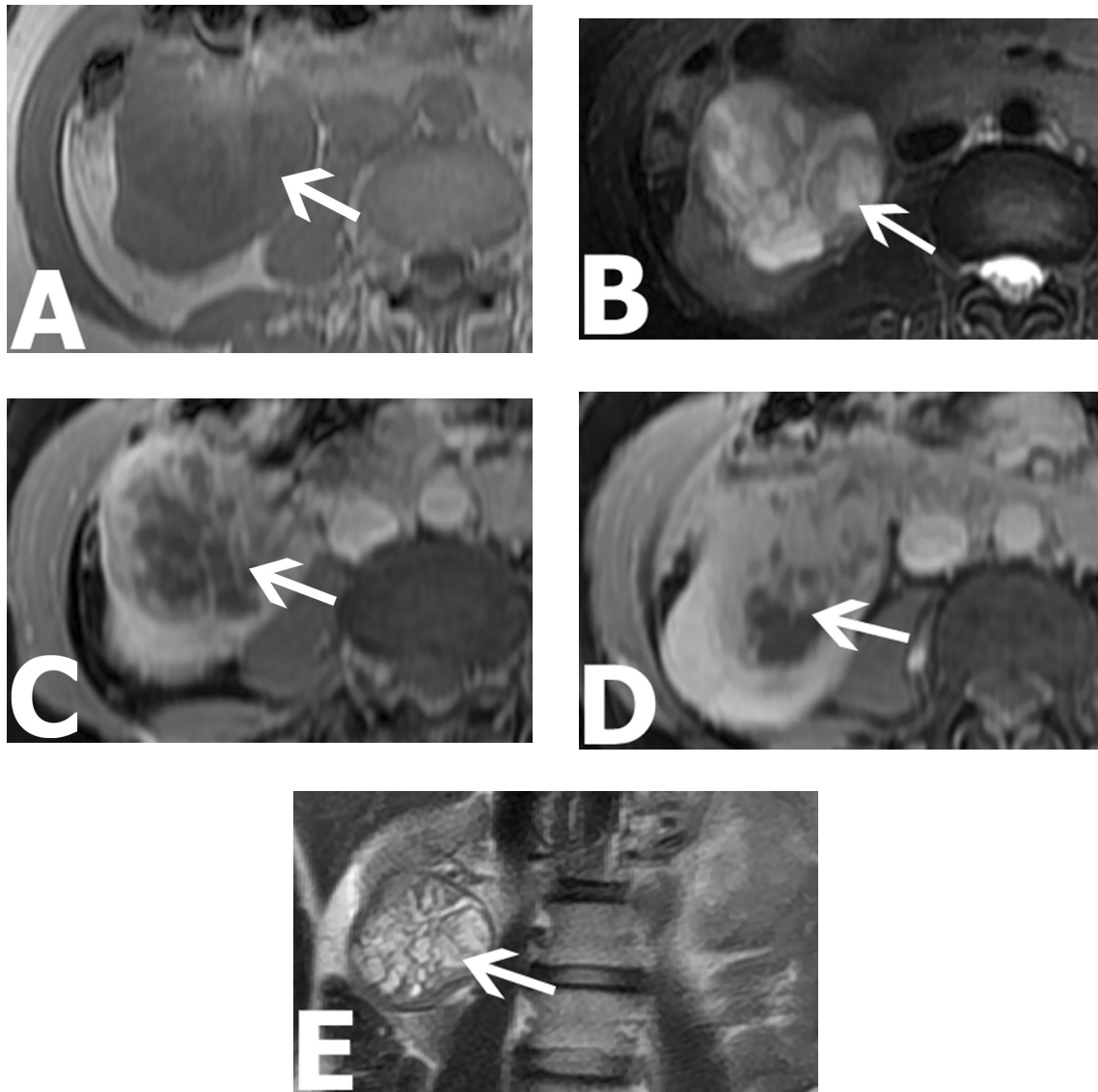


Figure 1 Axial in-phase T1 weighted gradient recalled echo (GRE) image (A), showing a heterogeneous mass lesion involving the medial portion of the lower pole of the right kidney (arrow), with a posterior isointense area and an anterior area of higher signal intensity, relative to skeletal muscle. Axial fat suppressed T2 (B) and coronal SSFSE T2 images (E) show marked heterogeneity and the 'triple sign': areas of low, intermediate and high signal intensity (arrows). Axial post-contrast GRE image (C and D) shows enhancement of the T2 hypo-intense areas (arrow).

negativity for S100, CK7, and epithelial membrane antigen (EMA). The diagnosis of primary synovial sarcoma of the kidney was made.

Discussion

The most common sarcoma of the kidney is leiomyosarcoma, accounting for 40–60% of reported cases. Primary SS of the kidney is a rare tumor^[4]. The first set of 15 cases reported was in 2000, by Argani^[4], with few other cases reported in the English literature. There have been no reports to date describing the MR imaging findings of primary SS of the kidney. The mean age of presentation

is 37.5 years. There is a slight male predominance, with a male-to-female ratio of 1.2. Furthermore, there seems to be a predilection for the right kidney, with a right-to-left ratio of 2.2.

The typical presentation of SS of the kidney is non-specific, and resembles that of other primary tumors of the kidney^[4]. Patients may complain of localized flank or back pain, hematuria, or a palpable abdominal mass. Imaging is often required for further evaluation.

For synovial sarcomas in general, the imaging modality of choice is magnetic resonance imaging (MRI), given its high sensitivity for soft tissue abnormalities. Although diagnostic for some soft tissue tumors, MR signal intensity characteristics are usually non-specific for SS^[1].

Most synovial sarcomas tend to be large masses, averaging 8 cm in size. The tumors have low invasion potential. The most common MR finding is an oval, well-defined nodular mass. On T1 weighted imaging, small lesions are typically homogenous, with a signal intensity similar to that of skeletal muscle^[5], while lesions larger than 5 cm often exhibit intermediate intensity signal and are heterogeneous secondary to hemorrhagic and necrotic areas^[11]. On T2 weighted imaging^[6,7], lesions are hyperintense, usually with intratumoral hemorrhage (73%) and cystic components (77%); 35–57% exhibit the triple sign, showing areas that are hyper-, iso-, and hypo-intense relative to skeletal muscle, representing hemorrhage/necrosis, septa/cellular elements, and calcified/fibrotic areas; however, this is also seen in other soft tissue tumors, namely malignant fibrous histiocytoma^[1,5]. MR imaging after intravenous contrast injection shows heterogeneous enhancement^[8].

Histologically, SS consists of round spindle cells with minimal cytoplasm and active mitotic figures. Cysts lined with epithelial cells containing eosinophilic cytoplasm are usually visible^[3]. These tumors are further divided into two subtypes, monophasic and biphasic, depending on the absence or presence of a well-developed glandular epithelium^[4]. The monophasic form is more common, and has a poorer prognosis. No radiographic distinction has been made between monophasic and biphasic tumors^[1].

Although, synovial sarcoma in any location is regarded as slow growing, it should be considered a systemic disease. Indeed, 25% have pulmonary metastases at the time of diagnosis. Despite wide surgical resection, local recurrence and metastatic disease are common, seen in more than 40%^[11]. There is no established treatment protocol for SS. However, SS, more than other sarcomas, are more often treated with neoadjuvant chemotherapy followed by surgical treatment^[9]. These tumors demonstrate moderate chemo sensitivity, with 50% response rates to ifosfamide-based or doxorubicin-based regimens^[4]. Studies of extremity SS have shown that ifosfamide-based chemotherapy reduces tumor volume by greater than 50% and improves disease-specific survival^[10]. In contrast, renal cell carcinoma is generally considered chemoresistant, although newer drug therapies such as sorafenib and sunitinib have led to some optimism^[11,12].

Prognosis of primary renal SS is unknown given the rarity of the diagnosis. Multiple studies of SS of the extremity have shown a strong association between

tumor size^[13], volume^[10], and histological grade^[14] to disease recurrence and patient mortality. Radiographic findings that had prognostic significance in SS were investigated^[15]. High grade SS is favored in tumors that exhibit cystic components, hemorrhage, and fluid levels, as well as the triple sign. Absence of calcification was also weakly associated with high-grade tumors.

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