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Case Report

Primary renal liposarcoma simulating angiomyolipoma ☆,☆☆

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ABSTRACT

Liposarcomas are infrequent malignant tumors of mesenchymal origin most commonly seen in the extremities. Although infrequent, these can develop as primary lesions in the soft tissue of the kidney, making them difficult to diagnose through imaging modalities alone. Primary renal liposarcomas are associated with poor prognoses, increasing the importance of timely and accurate diagnosis. In extremely rare instances, the tumor can arise directly from the fat in the epicenter of the kidney, disguised as an angiomyolipoma. In this article, we report the case of a 54-year-old female who was diagnosed with a well-differentiated liposarcoma of the kidney and underwent radical nephrectomy. Our objective is to evaluate unique radiological imaging findings and correlate with histopathological analysis to optimize diagnosis

Introduction

Primary renal liposarcomas are extremely rare tumors, representing 0.8% of all renal neoplasms and 2%-3% of malignant kidney cancers [1]. This tumor is diagnosed at a mean age of 50 and has been reported three times more frequently in women than men [1,2]. The infrequency of these tumors and the nonspecificity of clinical presentations pose a challenge in differentiating them from other renal tumors, such as angiomyolipomas or fat-containing renal cell carcinomas [3]. Renal liposarcomas typically have an asymptomatic onset and grow to sizes greater than 5 cm on average before causing symptoms [1]. Imaging modalities such as computed tomography (CT) and magnetic resonance imaging (MRI) are most effective in determining liposarcoma as a differential diagnosis,

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specifically through detecting the percentage of lipomatous content in the tumor; however, diagnostic confirmation can only be achieved through tissue examination [2].

Case presentation

A 54-year-old female with no significant medical history presented to the emergency department with right-sided abdominal pain and hematuria. She reported experiencing occasional right-sided mild abdominal pain that would resolve on its own. There was no associated nausea or vomiting, nor were there any changes in bowel movements. On review of systems, her weight had been stable, and she maintained a good appetite. She also recalled experiencing left-sided mild back pain while driving. The patient had a family history positive for breast cancer in her mother, and her father had a recent diagnosis of colon cancer. An ensuing CT scan with IV

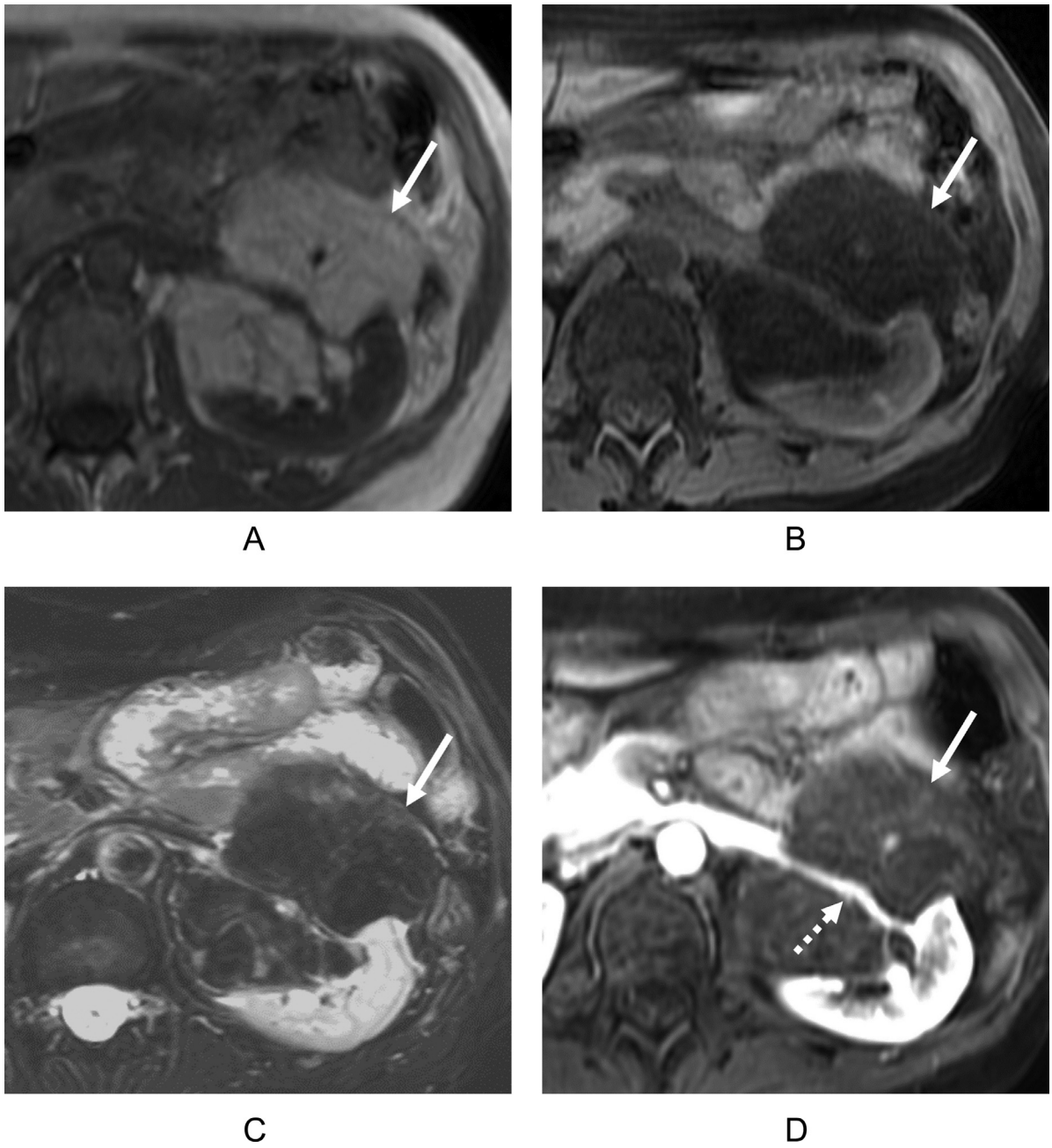

Fig. 1 – (A) Axial T1-weighted MR image showed a predominantly T1 hyperintense mass arising from the left renal hilum (arrow), with loss of signal on (B) axial T1-weighted fat-suppressed MR image (arrow), compatible with lipomatous tumor. (C) Axial T2-weighted fat-suppressed MR image showed a predominantly hypointense left renal hilar mass due to suppression of fat signal (arrow). A few T2 hyperintense thin internal septations were present. (D) Axial IV contrast-enhanced fat-suppressed T1-weighted MR image showed a heterogeneous enhancement of the mass arising from left renal hilum (arrow) with encasement and stretching of the left renal vasculature (dashed arrow).

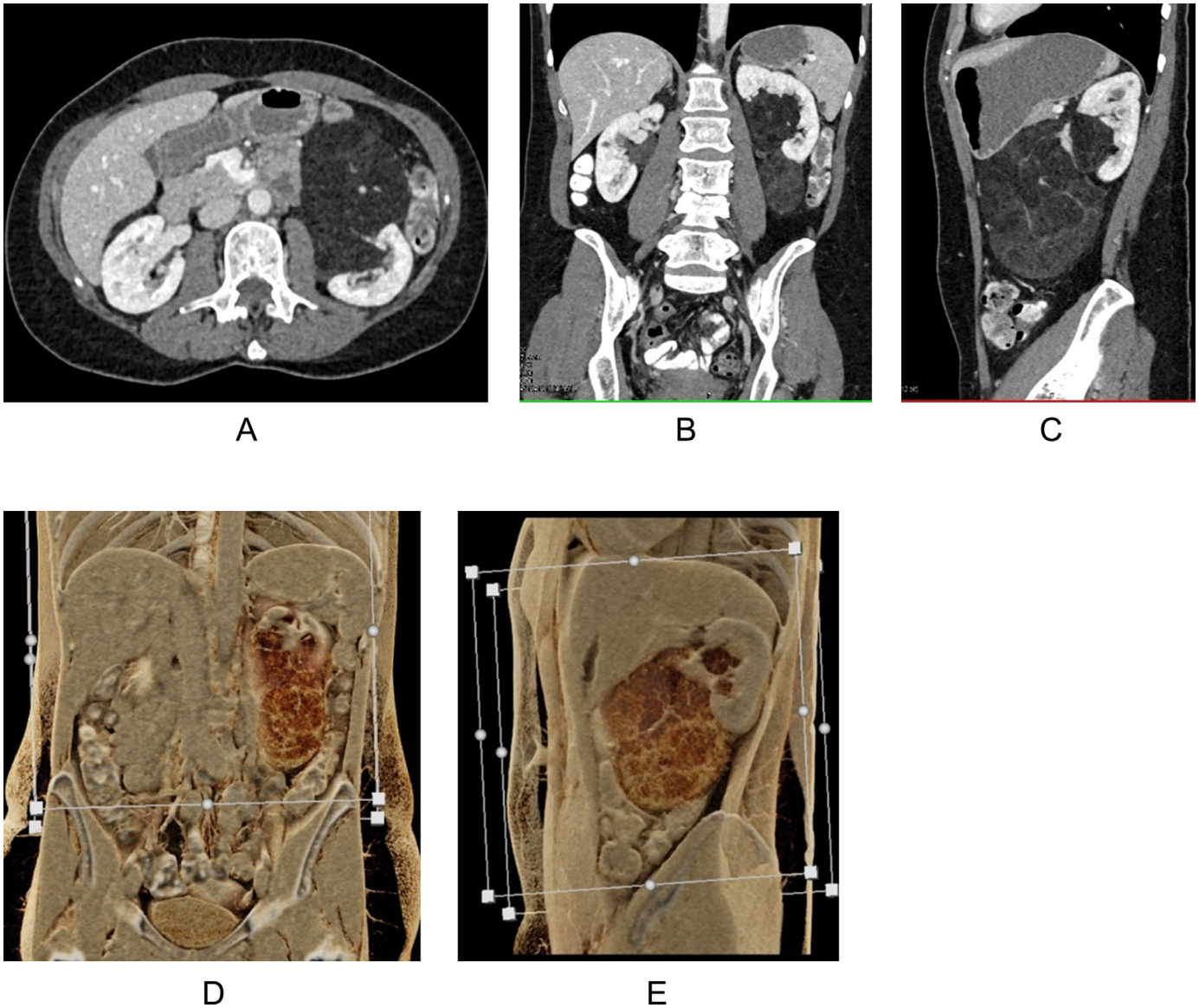

Fig. 2 – (A-C) Axial, coronal, and sagittal images define the extension of the tumor centrally into the kidney. Differential diagnosis included a renal AML versus a liposarcoma. (D and E) Cinematic rendering defines the extent of tumor, which appears to arise centrally in the kidney.

contrast of her abdomen and pelvis revealed a 6.7 × 9.4 × 12.0 cm fatty mass in the hilum of the left kidney extending anterior inferiorly within the retroperitoneum suggestive of an angiomyolipoma or a liposarcoma but was ultimately inconclusive. Further imaging was warranted to better characterize the soft tissue mass.

An MRI of the patient's abdomen (Figs. 1A-D) and pelvis showed a large fat-containing lesion involving the area of the left renal pelvis/hilum and infiltrating into the area of the collecting system/renal sinus fat, as well as extending inferiorly in the left retroperitoneum. This caused mild dilatation of the upper pole calyces secondary to compression of the renal pelvis/collecting system. The overall size of the lesion measured approximately 9.0 cm in the anterior/posterior direction and 8.6 cm transversely, with vertical dimensions of approximately 12.6 cm. In addition, it encased and displaced branches of the left renal artery and vein. The lesion also had internal septations and mild enhancement, more so inferiorly. The appearance favored a primary extrarenal liposarcoma invading/infiltrating the left renal sinus fat/renal pelvis rather than an angiomyolipoma extending exophytically.

Two weeks later, a CT scan (Figs. 2A-E) with IV contrast redemonstrated the large left-sided retroperitoneal mass extending from the full extent in anterior posterior dimension deviating structures. The mass measured 10.5 × 6.5 × 12 cm and was heterogeneous with predominantly low density, ranging from −15 to 60 Hounsfield units. The tumor encroached within the renal pelvis, and encasement of the left renal vein and left renal artery was observed as before. There was also a possible left upper pole calyceal dilatation that was noted. This large heterogeneous fat density mass that appeared to emerge from the left renal pelvis with a large intratumoral vessel was thought to be most consistent with an angiomyolipoma.

After imaging, the patient underwent an excision of the left retroperitoneal mass >10 cm as a whole with a left nephrectomy. The findings were a well-circumscribed mass arising from the left renal hilum separate from the left adrenal gland. Histopathological analysis confirmed a well-differentiated liposarcoma, FNCLCC (Fédération Nationale des Centres de Lutte Contre le Cancer) grade 1 of 3, forming a 12.5 cm mass with an epicenter in the renal sinus fat with retroperitoneal extension. RNA in situ hybridization for murine double-minute type 2 (MDM2) product amplification returned positive in part 1 (blocks 1E, 1N, 1U) but negative in part 2 (block 2D), supporting the liposarcoma diagnosis. The patient has been undergoing regular follow-up assessments with radiological imaging, clinical examination, and laboratory tests.

Discussion

Here, we review a case of primary renal liposarcoma, an extremely rare tumor emerging from lipoblasts of the kidney, accounting for 1%-2% of primary renal sarcomas [1]. Renal liposarcomas have been reported in patients ranging in age from 36 to 86 years old, with the majority being classified as well-differentiated tumors [1]. Clinical manifestations are nonspecific and most commonly include an abdominal mass with dimensions larger than 5 × 5 × 4 cm, pain, weight loss, and sometimes hematuria [4]. Differentiation of angiomyolipoma and liposarcoma based on patient demographics and tumor size is not possible due to similarities in characteristics.

Common differential diagnoses of renal liposarcomas include dedifferentiated pleomorphic sarcoma, pleomorphic liposarcoma, myxoid liposarcoma, and, most frequently, angiomyolipomas [1]. Angiomyolipomas appear to have similarities in terms of size and fat content to renal liposarcomas [1]. A retrospective study was conducted evaluating the characteristics to better differentiate angiomyolipomas from liposarcomas. On CT, angiomyolipomas may have linear vascularity, aneurysmal dilatation of intratumoral vessels, bridging vessel sign, hematoma, claw sign, and discrete intrarenal fatty tumors [5]. These aforementioned characteristics were not often seen in renal liposarcomas, rendering the distinction between the 2 possible [5]. Angiomyolipomas typically have an abundant fat content, leaving them easily identifiable on CT scans. However, this characteristic can also make it challenging to differentiate them from liposarcomas that arise specifically from renal sinus fat. In cases like ours, the similarities in radiographic features can be misleading, even for experienced radiologists. Ciccarello et al. in 2010 reported only 18 cases of liposarcoma arising from the renal capsule, and to the best of our knowledge, there have not been more significant reports [6]. Frequently seen imaging features of retroperitoneal liposarcomas include anterior displacement of the ipsilateral kidney, intramural calcification, and non–fat-attenuating enhancing intratumoral nodules [3].

In our case, the infiltrative/invasive nature of the mass seen on MRI raised concern for malignancy and suggested liposarcoma as a differential diagnosis. However, to reach a definitive diagnosis, a histopathologic examination was required. After the surgical removal of the mass via radical nephrectomy, pathologic results confirmed it as a well-differentiated liposarcoma. It is worth noting that the specimen tested positive for MDM2, a marker commonly associated with well-differentiated renal liposarcomas [7].

Conclusion

Renal liposarcomas are malignant cancers that develop from mesenchymal tissue and often grow to large sizes. Their asymptomatic nature and nonspecific symptoms cause delays in diagnosis, and the prognosis is associated with tumor characteristics such as differentiation, size, histologic type, and tumor stage, along with the kind of surgical resection that occurs [1,4]. Due to the malignant nature of these tumors, primary renal liposarcomas should be treated with radical surgical resection as quickly as possible [5]. In our case, although the patient was initially thought to have an angiomyolipoma, the mass was going to be resected due to the sheer size. The use of chemotherapy and radiotherapy has not been frequently reported, and the results are incongruent [1].

Patient consent

The patient reported in the manuscript signed the informed consent/authorization for participation in research, which includes the permission to use data collected in future research projects such as the presented case details and images used in this manuscript.

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