J Med Ultrasonics (2016) 43:427–430
DOI 10.1007/s10396-016-0705-1

CASE REPORT

Contrast-enhanced ultrasound in diagnosis of epithelioid renal angiomyolipoma with renal vein and inferior vena cava extension

Tingting Qiu[1] · Wenwu Ling[1] · Qiang Lu[1] · Changli Lu[1] · Yan Luo[1]

Received: 17 December 2015 / Accepted: 1 February 2016 / Published online: 9 March 2016

Abstract Epithelioid angiomyolipoma is considered to be a rare variant of angiomyolipoma and potentially malignant. Reported cases of renal epithelioid angiomyolipoma with extension into the renal vein and inferior vena cava are even rarer. Once it occurs, it is difficult to differentiate from renal cell carcinoma on imaging. In addition, very few of the reported cases presented contrast-enhanced ultrasound images. Here, we report a case of a renal epithelioid angiomyolipoma extending into the renal vein and inferior vena cava along with its contrast-enhanced ultrasonic information. Contrast-enhanced ultrasound provided valuable information for diagnosis in this case.

Keywords Contrast-enhanced ultrasound · Epithelioid angiomyolipoma · Renal vein · Inferior vena cava

Introduction

Epithelioid angiomyolipomas (EAMLs) are closely associated with angiomyolipomas (AMLs). AMLs are always considered benign, while EAMLs are thought to be potentially malignant. To date, only a small number of renal EAMLs with vascular involvement or distant metastases have been reported [1–3]. Most of the reported cases presented CT or MRI results, while few of them demonstrated detailed ultrasonic or contrast-enhanced ultrasonic information. Here, we report a case of a renal epithelioid angiomyolipoma with vascular extension. Contrast-enhanced ultrasound (CEUS) was applied in this case and was helpful for diagnosis.

Case presentation

A 44-year-old woman was incidentally diagnosed with a right renal mass on gray-scale ultrasound. Ultrasonography revealed a 5.0 × 3.2-cm hyperechogenic mass occupying the inferior pole of the right kidney (Fig. 1a). The mass engulfed part of the renal medulla and was not well-circumscribed. Extension of the mass into the right renal vein (RRV) and inferior vena cava (IVC) was about 6.0 cm long (Fig. 1e). The patient then underwent CEUS for further diagnosis. After acquisition of informed consent, bolus injection of 1.2 ml of a contrast agent (SonoVue) suspension followed by a 5-ml saline flush was performed through the cubital vein. During the corticomedullary phase of CEUS, the mass was heterogeneously hyperenhanced compared to adjacent renal parenchyma (Fig. 1b). In the nephrographic and excretory phases, the mass was isoenhanced (Fig. 1c, d). The mass in the RRV and IVC was hypoenhanced the whole time (Fig. 1f). Based on the contrast pattern and the involvement of vessels, AML was suspected, but renal cell carcinoma (RCC) could not be ruled out. Contrast-enhanced computed tomography (CECT) revealed a mass of irregular shape at the inferior pole of the right kidney. The mass was heterogeneously hyperenhanced in the corticomedullary phase and isoenhanced in the late phase (Fig. 2a, b). A filling defect was detected in the RRV and IVC (Fig. 2b, c). After CECT, diagnosis of renal tumor with vascular involvement was made. Malignancy was still undetermined. The patient did

✉ Yan Luo
luoyanddoc@163.com

[1] Department of Ultrasound, West China Hospital of Sichuan University, 37 Guoxue Lane, Chengdu 610041, Sichuan, China

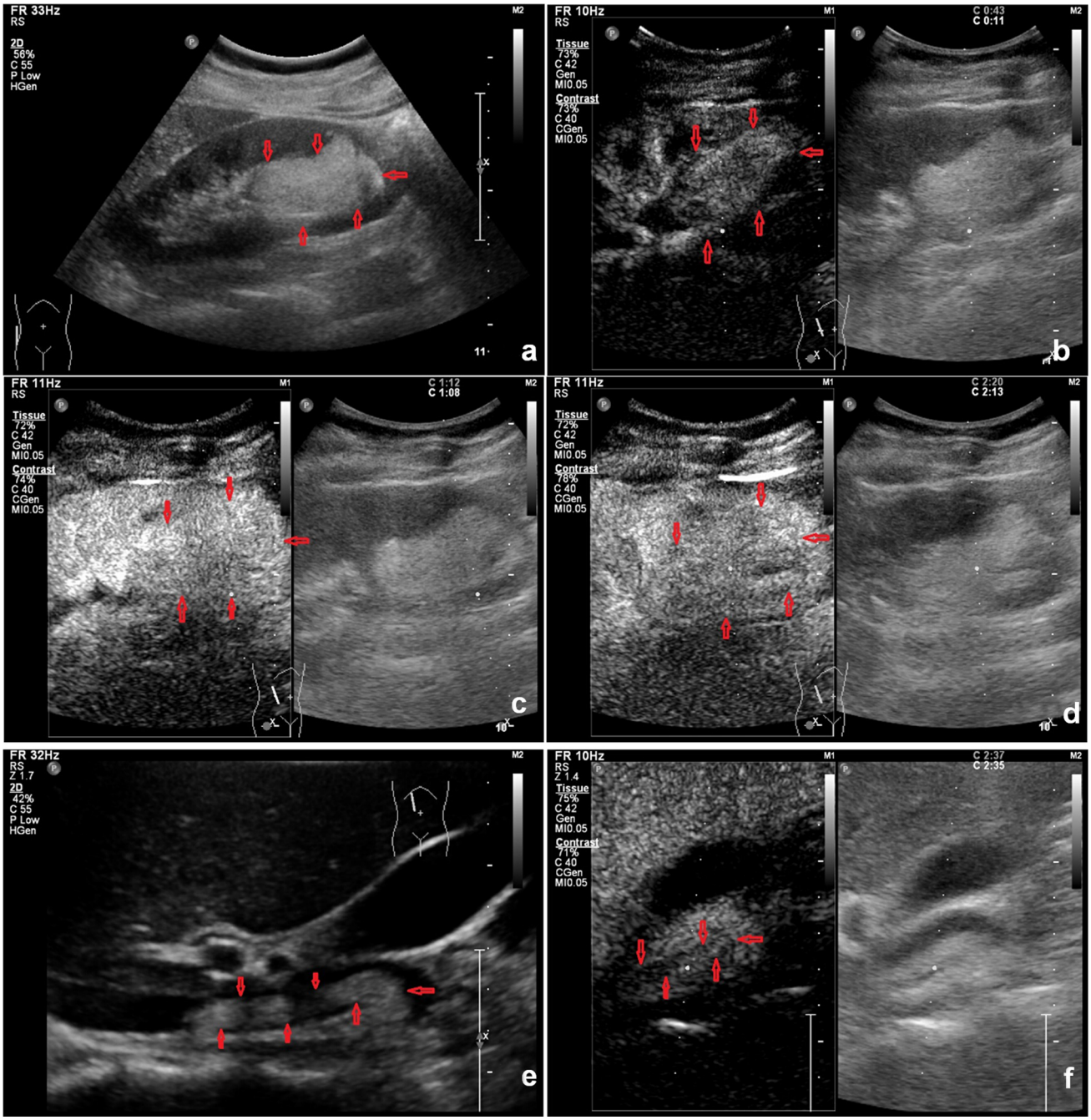

Fig. 1 B-mode ultrasound images. **a** A 5.0 × 3.2-cm hyperechogenic mass occupying the inferior pole of the right kidney. **e** Extension of the mass into the RRV and IVC. Contrast-enhanced ultrasound images. **b** The mass was heterogeneously hyperenhanced compared to adjacent renal parenchyma during the corticomedullary phase. **c**, **d** The mass was isoenhanced in the nephrographic and excretory phases. **f** Extension of the renal mass in the RRV and IVC was hypoenhanced the whole time

not undergo MRI. Right radical nephrectomy with IVC thrombectomy was performed successfully. During the 5-h-surgery, the renal tumor was found at the inferior pole of the right kidney, 5.0 × 3.0 cm in size, projecting outside of the renal contour. A yellow and soft thrombus was present in the RRV and IVC with no adhesion to the vessel wall. About 500 ml of blood was lost during surgery. After surgery, both the right renal mass and the thrombus in the RRV and IVC were confirmed as epithelioid angiomyolipoma (Fig. 3a, b). Immunohistochemical staining for the antibodies to CD31, CD34, and D2–40 was all positive (Fig. 3c).

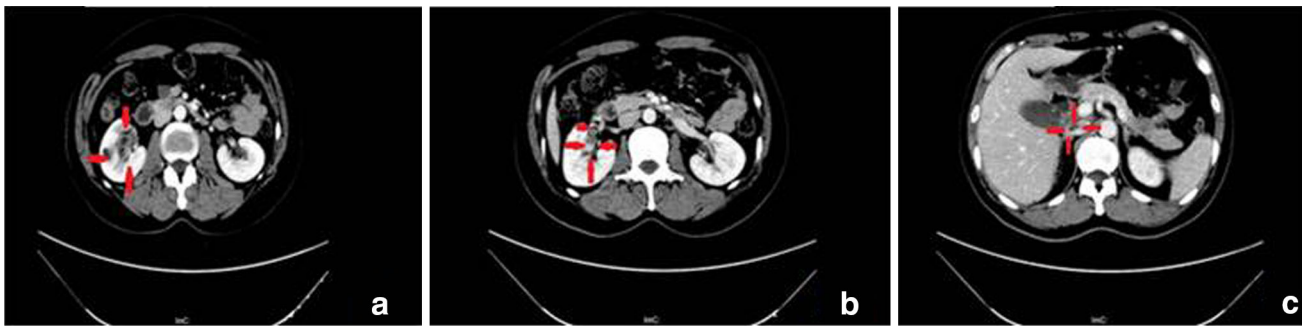

Fig. 2 Contrast-enhanced computed tomography images. **a**, **b** The mass was heterogeneously hyperenhanced in the corticomedullary phase and isoenhanced in the late phase. **b**, **c** A filling defect was detected in the RRV (**b**) and IVC (**c**), revealing a tumor thrombus in the vein

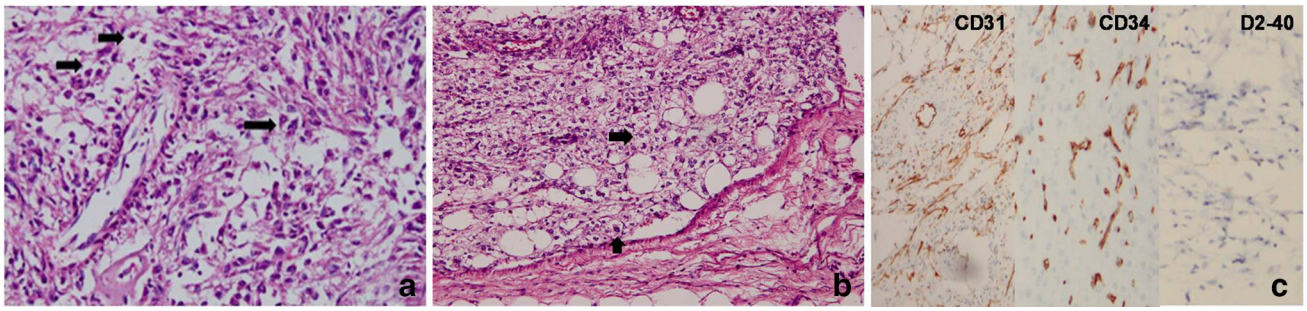

Fig. 3 Epithelioid angiomyolipoma. Photomicrograph **a** and **b** (hematoxylin–eosin staining; magnification ×200). Both the renal mass (**a**) and its extension (**b**) in the vein demonstrated sheets of epithelioid cells with moderate nuclear atypia or multi-nuclei, which characterize this tumor. **c** Immunohistochemical staining (magnification ×200). CD31, CD34, and D2–40 were all positive, indicating vascular and lymphatic endothelial cells of this tumor

Discussion

From the perspective of pathology, epithelioid angiomyolipoma (EAML) is defined as a potentially malignant mesenchymal neoplasm characterized by proliferation of predominantly epithelioid cells [4]. Nevertheless, the percentage of epithelioid cells required for diagnosing EAML is still under debate, ranging from 5 to 100 % [5, 6].

As for imaging diagnosis, conventional ultrasound (CUS) is the first choice for detecting renal masses as it is inexpensive, noninvasive, non-ionizing, and readily available. However, CUS has limited ability in differentiating RCCs and AMLs [7]. Compared to CUS, CEUS has the advantage of displaying vascular perfusion, which is meaningful for the determination of tumor malignancy. Studies have shown that AMLs tend to be more homogeneously enhanced than clear cell RCCs [8, 9]. But, this feature is limited to renal masses larger than 3 cm, because RCCs smaller than 3 cm tend to have a low incidence of necrosis. Hence, they can be as homogeneously enhanced as AMLs [7, 10]. EAMLs are usually enhanced early on CEUS, but RCCs are always enhanced even earlier [11]. Washing out of ultrasound contrast agents in AMLs is prolonged compared to clear cell RCCs, which may be another characteristic for differentiation [8, 11]. CEUS is also able to demonstrate peritumoral rim-like enhancement. This type of enhancement may be related to peritumoral compression and inflammation due to malignant tumor growth [7, 8, 12]. Overall, CEUS has been shown to outperform CUS with significantly higher accuracy in differentiating small renal masses [7, 13]. In our case, the pathologically confirmed EAML presented heterogeneously early hyperenhancement in the corticomedullary phase and isoenhancement in the nephrographic and excretory phases on CEUS. Therefore, it is consistent with previous study results and the CECT results in the present case [8, 9, 11]. Considering radiation exposure and the higher cost of CECT, CEUS is obviously the better choice given its diagnostic ability relative to CECT. Additionally, Andre Ignee et al. [13] studied 12 patients with renal masses and vascular involvement. They found that CEUS was comparable to CECT in staging of vein invasion. And only CEUS, but not CDI or CECT could display vascularization of thrombus in the vein. With vascular involvement, it is hard to rule out RCCs completely, because other types of RCCs including papillary and chromophobe RCCs may display a similar enhancement pattern on CEUS [14]. To our knowledge, there have been almost no studies

comparing CEUS characteristics between EAMLs and AMLs, with the exception of some individual reports, because EAMLs are more infrequently seen than AMLs. On the other hand, CEUS has not been used as prevalently as CT or CECT to date. Tsai et al. summarized 20 cases of CT findings of EAMLs. They reported no fat or calcification and "good enhancement" in the early contrast-enhanced phase, which are characteristics that might differentiate EAMLs from AMLs on CT images [15].

Conclusion

We presented a rare case of a renal EAML with renal vein and inferior vena cava extension. In this case, CEUS provided valuable diagnostic information.

Acknowledgments This work was supported by the National Natural Science Foundation of China, No. 0040205401927.

Compliance with ethical standards

Conflict of interest There are no financial or other relations that could lead to a conflict of interest.

Ethical statements All procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with the Helsinki Declaration of 1975, as revised in 2008. Informed consent was obtained from the patient for being included in this case.

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