A young female case of primary renal carcinoid tumor mimicking fat-poor angiomyolipoma

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Abstract

Renal mass biopsy is useful for the pathological diagnosis of a small renal mass difficult to distinguish fat-poor angiomyolipoma from renal cell carcinoma radiologically. Here, we report a young female case of a small renal mass suspected as fat-poor angiomyolipoma in imaging studies. The patient received a renal mass biopsy to obtain the correct pathological information. Unexpectedly, the pathological diagnosis was neuroendocrine tumor. She, finally, underwent a right radical nephrectomy as a curative treatment. This case indicates that the radiological findings of primary renal carcinoid tumor can be similar to those of fat-poor angiomyolipoma.

Keywords Angiomyolipoma · Neuroendocrine tumors · Biopsy

Introduction

In the management of small renal mass (SRM), the preoperative differential diagnosis of fat-poor angiomyolipoma (AML) from renal cell carcinoma (RCC) is still a clinical challenge despite recent advances in imaging studies. Here, we report a young female case with a SRM clinically suspected as fat-poor AML. In this case, renal mass biopsy (RMB) resulted in an unexpected pathological diagnosis of neuroendocrine tumor. We further highlight the radiological findings of fat-poor AML and primary renal carcinoid tumor and discuss their similarities in imaging studies.

Case report

A 43-year-old female patient presented with an incidental small right renal mass on ultrasound at medical checkup. She had no medical history and no symptoms. On ultrasound, the mass appeared heterogeneously hyperechoic (Fig. 1a). CT showed a 3-cm-large right renal mass which was homogeneously enhanced but less than the renal cortex in corticomedullary phase (Fig. 1b-d). MRI showed a low-signal

intensity mass without a pseudocapsule on T2-weighted image (Fig. 1e). There was no fat component in the mass on CT and MRI. Thus, we clinically diagnosed with fat-poor AML. To exclude the possibility of RCC, we conducted a RMB. Unexpectedly, the pathological examination of the biopsy specimen revealed neuroendocrine tumor. Preoperative positron emission tomography showed no lymph-node and/or distant metastasis. Because the tumor was ill-defined on ultrasound (Fig. 1a), we were afraid of the risk of positive surgical margins at partial nephrectomy. Thus, we performed a right minimum incision endoscopic radical nephrectomy to achieve the complete resection of the tumor [1]. Intraoperative bleeding was 14 ml, and the total duration of the surgery was 115 min. No complications occurred postoperatively, and she was discharged on day 8. Macroscopically, the tumor was a solid, grayish-yellow mass of 3 cm in size (Fig. 2a). On microscopic examination, tumor cells with eosinophilic cytoplasm and uniform round nuclei were arranged in a nested and alveolar pattern (Fig. 2b). Immunohistologically, the tumor cells were positive for chromogranin A (Fig. 2c), synaptophysin (Fig. 2d), and somatostatin receptor type 2a (Fig. 2e). The mitotic count and the Ki-67 index of the tumor were 5 mitoses/10 high-power fields and 10%, respectively. Thus, the tumor was pathologically diagnosed with neuroendocrine tumor, grade 2. The patient is alive without recurrence 3 months after the surgery.

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Fig. 1 Ultrasound, CT, and MRI images of the tumor. The tumor appeared heterogeneously hyperechoic on ultrasound (**a**, white arrowhead) and was homogeneously enhanced but less than the renal cortex in corticomedullary phase on CT (**b** unenhanced, **c** corticomedullary phase, and **d** nephrographic phase). MRI showed a low-signal intensity mass without a pseudocapsule on T2-weighted image (**e**). There was no fat component in the mass on CT and MRI

Discussion

In the present case, the tumor appeared to be fat-poor AML in imaging studies. The tumor was hyperechoic on ultrasound. On CT, it was homogeneously enhanced but less than the renal cortex in corticomedullary phase. MRI showed a low-signal intensity mass without a pseudocapsule on T2-weighted image. No fat component was observed in the tumor. All these findings were compatible with fat-poor AML. Therefore, we performed an RMB to obtain the pathological diagnosis of AML and exclude the possibility of RCC. Interestingly, we obtained the unexpected pathological diagnosis of neuroendocrine tumor.

AML is a benign tumor whose incidence is relatively high in young females [2]. AML typically consists of

Fig. 2 Macroscopic and microscopic findings of the resected tumor. Macroscopically, the tumor was a solid, grayish-yellow mass (**a**). Microscopically, tumor cells with uniform round nuclei were arranged in a nested and alveolar pattern (**b** $\times 10$). Immunohistochemically, the tumor cells were positive for chromogranin A (**c** $\times 20$), synaptophysin (**d** $\times 20$), and somatostatin receptor type 2a (**e** $\times 20$)

dysmorphic blood vessels, smooth muscle components, and mature adipose tissue. AML is usually diagnosed based on the fat component in the mass on CT [3]. However, fat-poor AML is sometimes difficult to diagnose, because its findings are radiologically similar to those of RCC [4]. Tanaka et al. proposed a stepwise algorithm for the differential diagnosis of fat-poor AML and RCC [5]. Their algorithm included (1) female age < 50 years, (2) high attenuation on unenhanced CT, (3) less enhancement in corticomedullary phase of enhanced CT, (4) low-signal

intensity on T2-weighted MRI, and (5) the absence of pseudocapsule on T2-weighted MRI. According to their algorithm, the present case was classified as the high AML-probability group.

Primary renal carcinoid tumor is very rare. Carcinoid tumors represent well-differentiated neuroendocrine tumors arising in the gastrointestinal tract, lungs, or rare primary sites, such as the kidneys or ovaries [6]. According to the literature review in 2006, only 56 reported cases of primary renal carcinoid tumor were found by an extensive search [7]. Approximately half of the patients had metastatic disease despite its indolent course. Patients who achieved no evidence of disease after treatment showed excellent prognosis [7]. A paucity of data exist on imaging findings of primary renal carcinoid tumor. Omiyale et al. reported that renal carcinoid tumor showed a hyperechoic mass with an incomplete hypoechoic thin rim and calcifications on ultrasound [8]. Moreover, they reported that typical CT findings of renal carcinoid tumors were minimal or poor contrast enhancement on CT. Considering that the present case showed no calcification, primary renal carcinoid tumor without calcification may have similar imaging features to fat-poor AML.

RMB is useful to obtain pathological information for the diagnosis of SRM [9]. In the present case, RMB contributed to the pathological diagnosis of neuroendocrine tumor. Thus, RMB can be recommended to obtain a correct pathological diagnosis in a case with a SRM suspected as fat-poor AML in imaging studies. However, the most disastrous complication of RMB is tumor seeding of the biopsy tract. The incidence of tumor seeding by RMB is extremely rare in RCC [10]. Because the rate of tumor seeding is unknown in neuroendocrine tumor, careful follow-up is mandatory for the present case.

In conclusion, the radiological findings of primary renal carcinoid tumor can be similar to those of fat-poor AML. The present case emphasizes the diagnostic role of RMB in a case with a SRM suspected as fat-poor AML in imaging studies.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

Ethical approval All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

References

1. Iimura Y, Kihara K, Saito K et al (2008) Oncological outcome of minimum incision endoscopic radical nephrectomy for pathologically organ confined renal cell carcinoma. *Int J Urol* 15:44–47
2. Fujii Y, Komai Y, Saito K et al (2008) Incidence of benign pathologic lesions at partial nephrectomy for presumed RCC renal masses: Japanese dual-center experience with 176 consecutive patients. *Urology* 72:598–602
3. Jinzaki M, Silverman SG, Akita H et al (2014) Renal angiomyolipoma: a radiological classification and update on recent developments in diagnosis and management. *Abdom Imaging* 39:588–604
4. Schieda N, Hodgdon T, El-Khodary M et al (2014) Unenhanced CT for the diagnosis of minimal-fat renal angiomyolipoma. *Am J Roentgenol* 203:1236–1241
5. Tanaka H, Fujii Y, Tanaka H et al (2017) Stepwise algorithm using computed tomography and magnetic resonance imaging for diagnosis of fat-poor angiomyolipoma in small renal masses: Development and external validation. *Int J Urol* 24:511–517
6. Kulke MH, Mayer RJ (1999) Carcinoid tumors. *N Engl J Med* 340:858
7. Romero FR, Rais-Bahrami S, Permpongsol S et al (2006) Primary carcinoid tumors of the kidney. *J Urol* 176:2359–2366
8. Omiyale AO, Venyo AK (2013) Primary carcinoid tumour of the kidney: a review of the literature. *Adv Urol* 2013:579396
9. Lim A, O'Neil B, Heilbrun ME et al (2012) The contemporary role of renal mass biopsy in the management of small renal tumors. *Front Oncol* 2:106
10. Blute ML, Drewry A, Abel EJ (2015) Percutaneous biopsy for risk stratification of renal masses. *Ther Adv Urol* 7:265–274