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Myelolipoma of renal sinus in a patient with end-stage renal disease: A rare differential diagnosis in renal neoplasms

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Abstract

Myelolipoma is an uncommon benign neoplasm composed of mature adipose tissue and hematopoietic elements. Myelolipoma occurs most frequently in the adrenal gland but occasionally can be seen in extra-adrenal locations such as lung, liver, retroperitoneum, mediastinum, and kidney. We report a case of extra-adrenal myelolipoma presented as a localized asymptomatic mass in the renal sinus of left native kidney of a 60-year-old woman. The patient was being investigated for end-stage renal disease status post renal transplant. Histologic examination of the lesion showed classic features for myelolipoma. Our case illustrates the awareness of this entity in unusual location and the appropriate use of diagnostic modalities to prevent unnecessary surgical interventions particularly in patients with major co-morbidity in the future.

Keywords

Myelolipoma, extra-adrenal, renal sinus, kidney, retroperitoneum

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Introduction

Myelolipomas are benign mesenchymal tumors composed of mixture of mature adipose tissue and hematopoietic elements.[1] Although myelolipomas most commonly occur in adrenal glands but they have been occasionally found in extra-adrenal location. More than half of extra-adrenal myelolipomas have been localized to the presacral region.[2,3] Other uncommon reported locations include pelvis, thorax, retroperitoneal space, and perirenal tissue.[4] Widespread use of imaging techniques has led to increased detection of extra-adrenal myelolipomas. The majority of patients are asymptomatic at the time of diagnosis and lesions are discovered incidentally on imaging for other medical problems.

Case presentation

We present a 60-year-old female with end-stage renal disease and significant past medical history of diabetic nephropathy and chronic hemodialysis. Patient underwent deceased donor renal transplant (DDRT) in 2018. Her transplant was initially complicated by delayed graft function. Two consecutive renal biopsies showed mild acute tubular injury without any evidence of antibody-mediated or cell-mediated rejection. Patient continued the hemodialysis for a few more months until graft function returned to normal. Her creatinine level changed from 1.89 mg/dL to current baseline level of 1.1–1.2 mg/dL. In 2019, an incidental mass was found in her left native kidney during routine post renal transplant workup. Abdominal ultrasound revealed an echogenic area with increased vascularity in the mid pole of left native kidney most likely representing prominent renal sinus fat based on sonogram interpretation. An abdominal magnetic resonance imaging (MRI) with and without contrast showed a subtle T2 hypointense, T1 intermediate lesion at the interpolar renal hilum measuring up to 2.6 cm demonstrating mild early enhancement, diffusion restriction, and delayed washout, concerning for renal cell carcinoma or less likely urothelial carcinoma of renal pelvis (Figure 1(a)). CT urogram showed a questionable heterogeneous enhancement within the left renal pelvis (Figure 1(b)). Given the concern of urothelial carcinoma, diagnostic ureteroscopy was

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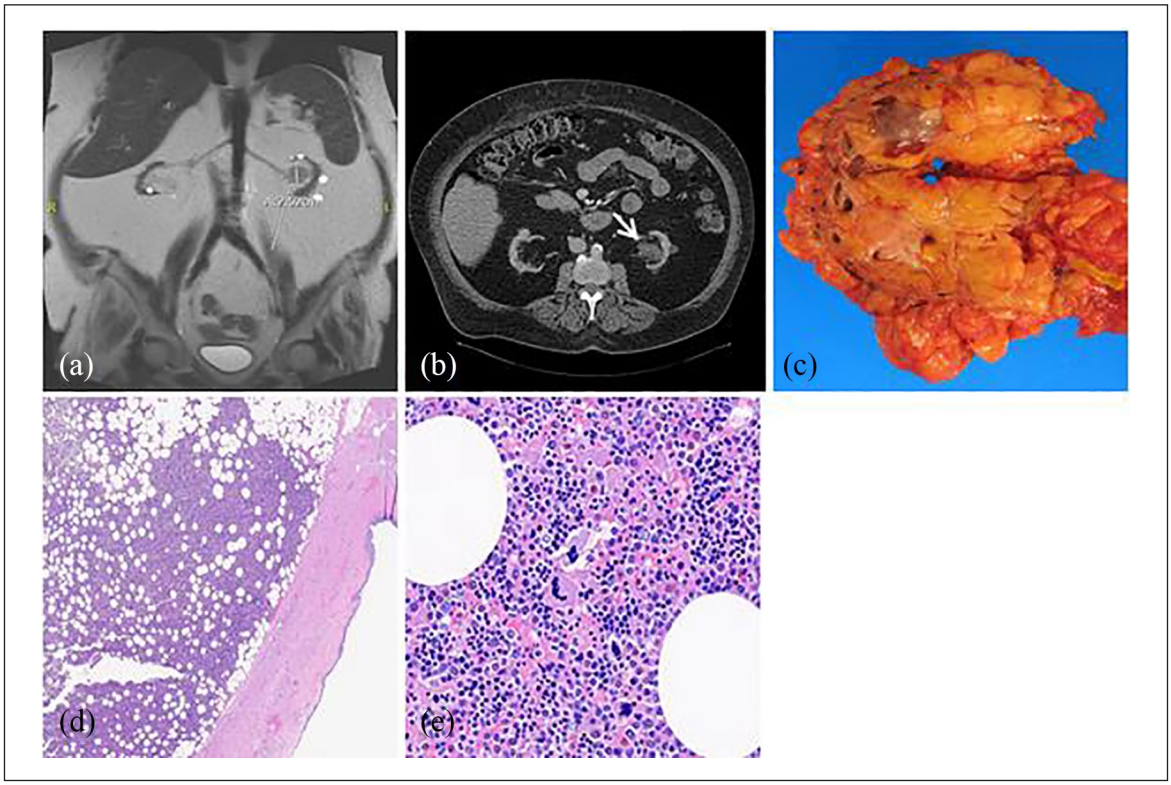

Figure 1. (a) MRI shows subtle T2 hypodense, T1 intermediate lesion at the interpolar renal hilum of left native kidney measuring up to 2.6 cm. (b) CT urogram shows a questionable heterogeneous enhancement within the left native renal pelvis (arrow). (c) Well-circumscribed mass in renal sinus in the background of left atrophic native kidney. (d) Low magnification (20×) shows mature adipose tissue and hematopoietic elements. (e) Higher magnification (100×) shows normal hematopoietic elements, such as megakaryocytic, erythropoietic, and granulopoietic cell lineages dispersed within adipose tissue.

performed but did not show any mass in the renal pelvis. Due to anatomic location of the mass and major imaging and clinical concern for malignant process, like renal cell carcinoma, endoscopic ultrasound-guided needle biopsy of mass was performed. Histopathological examination was not conclusive for definite pathologic diagnosis due to inadequate tumor sampling. Considering high risk of renal cell carcinoma in patient due to years on dialysis and chronic immunosuppression, the patient subsequently underwent laparoscopic nephrectomy of native kidney. Gross pathologic examination of the resected native kidney revealed an atrophic kidney with a well-circumscribed, non-encapsulated, tan-brown mass in the renal sinus adipose tissue measuring 3.0 cm × 2.5 cm × 1.7 cm (Figure 1(c)). Histologic examination of the mass showed mature adipose tissue admixed with benign hematopoietic elements representing all three hematopoietic cell lineages diagnostic for extra-adrenal myelolipoma (Figure 1(d) and (e)). Patient had an uneventful postoperative and recovery phase.

Discussion

Myelolipomas are benign mesenchymal tumors composed of mature adipose tissue intermixed with mature hematopoietic elements. Hematopoietic cells represent a variable percentage of all myeloid, erythroid, and megakaryocyte lineages.[1] Edgar von Gierke first described this entity in the adrenal gland in 1905, but it was finally named myelolipoma in 1929 by Charles Oberling.[5,6] The adrenal gland is the most common site. Myelolipoma is the second most common adrenal cortical tumor. It accounts for 2.6% of all primary adrenal tumors.[2] However, extra-adrenal myelolipomas are exceedingly rare and constitute less than 15% of all myelolipomas.[3]

Extra-adrenal myelolipomas may occur in the presacral region, retroperitoneum, pelvis, thorax, mediastinum, stomach, liver, and even in the thyroid glands.[4] Extra-adrenal myelolipomas rarely occur in the kidney or perirenal adipose tissue with only eight cases have been reported in English literatures within the last three decades. Both adrenal and extra-adrenal myelolipomas occur more commonly in older patients with female predominance. They are mostly asymptomatic and usually discover incidentally during a radiological investigation of an unrelated condition. Abdominal or flank pain are most common complains in symptomatic patients which can be due to intratumoral or peritumoral hemorrhage, infarction, or mechanical compression of the tumor on adjacent structures.[7]

Although the etiology of myelolipoma is not completely understood, a widely accepted theory holds that myelolipomas may be remnants of fetal bone marrow. Other hypotheses include the possibility that they arise as a result of embolism of bone marrow cells or hyperplasia of heterotopic reticulum cells.[8,9] In the embryonic stage of fetal development, hematopoiesis occurs in the peritoneal connective tissue but regresses with the development of other hematopoietic centers. Activation of retroperitoneal hematopoietic centers has been seen in many infectious conditions, which supports a similar causative mechanism for adrenal or extra-adrenal myelolipoma.[10–12]

Due to presence of mature adipose tissue in myelolipoma, radiologic techniques such as CT scan and MRI can precisely diagnose myelolipoma in the adrenal gland.[13] However, extra-adrenal myelolipomas and fat-poor myelolipomas are more challenging to diagnose preoperatively because they are easily confused with some other lesions. Differential diagnoses of a fat-containing retroperitoneal mass include retroperitoneal liposarcoma, adrenal or extra-adrenal myelolipoma, angiomyolipoma, and retroperitoneal teratoma. If a definite diagnosis is needed, image-guided fine-needle biopsy is indicated.[14,15]

Grossly extra-adrenal myelolipoma is usually a solitary mass. The mean tumor size is 3–5 cm, and the largest reported tumor measured 27 cm.[14] The tumor is well-demarcated, round to oval. The tumor cut surface has a heterogeneous yellow-to-red appearance depending on the proportions of the fat and hematopoietic components.[7] Microscopically, myelolipomas are composed of mature adipose tissue and variable percentage of bone marrow elements. The three hematopoietic cell lineages include myeloid, erythroid, and megakaryocyte. Areas of hemorrhage, infarction, calcification, or osseous metaplasia are occasionally noted.[7] Molecular testing or immunohistochemical staining does not have any diagnostic or clinical value in myelolipomas.

Extra-adrenal myelolipomas are benign neoplasms with excellent long-term prognosis.[16] However, they may enlarge and have a risk of bleeding. Malignant transformation has not been reported to date. Some studies with long-term follow-up (3–62 months) have shown stable clinical course.[17] In most institutions, small asymptomatic lesions will be managed conservatively with radiology follow-up due to small potential of hemorrhage and rapid growth. In symptomatic patients with an uncertain diagnosis or enlarging mass, surgical intervention is warranted.

However, in patients with other co-morbidities due to difficulty in major surgery and higher risk of side effects, pros and cons of surgical intervention should be precisely evaluated. In the meantime, having definite diagnosis with proper use and interpretation of diagnostic modalities such as MRI, CT scan, and image-guided needle biopsy can prevent unnecessary surgical interventions. Unfortunately, due to anatomic location of the mass in our patient, the conventional transcutaneous image-guided needle biopsy was not possible. Patient underwent endoscopic ultrasound-guided needle biopsy, which given its limitation, the mass was not adequately sampled; therefore, the final pathology result was not conclusive for a definite diagnosis.

Conclusion

Extra-adrenal myelolipomas, particularly those arising in perirenal soft tissue, renal hilum, and renal sinus are exceedingly rare. Only about eight cases of renal or perirenal myelolipomas have been reported in English literatures within the last three decades. An awareness of renal myelolipoma occurrence should expand our differential diagnosis in renal and perirenal masses. The differential diagnoses to consider for such lesions include but not limited to myelolipoma, angiomyolipoma, lipoma, liposarcoma, urothelial carcinoma, renal cell carcinoma, and metastatic tumors. Extra-adrenal myelolipoma can be managed conservatively based on the degree of certainty of primary diagnosis. Appropriate use of diagnostic imaging modalities in combination with image-guided needle biopsy can prevent a major surgical intervention particularly in patients with major co-morbidities as in our patient. Nevertheless, improper use or interpretation of above diagnostic methods can hinder their benefits in the prevention of a major surgical intervention for patients.

Declaration of conflicting interests

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Ethical approval

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