Case Report

Rare huge bilateral adrenal myelolipoma confused with retroperitoneal liposarcoma

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Abbreviations & Acronyms
ACTH = adrenocorticotropic hormone
CT = computed tomography
FDG-PET = fluorodeoxyglucose positron emission tomography
HU = Hounsfield units
MRI = magnetic resonance imaging

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Introduction: Adrenal myelolipomas are uncommon benign adrenal tumors, which mostly occur unilaterally. We describe a rare case of giant bilateral adrenal myelolipoma mistaken for retroperitoneal liposarcoma.

Case presentation: A 49-year-old man developed fever, left flank pain, and a large mass in his left abdomen. Plain computed tomography showed a 23-cm mass with fat-containing components in the retroperitoneal cavity, suggestive of retroperitoneal liposarcoma. A similar 9-cm mass was observed in the right adrenal gland, suggesting adrenal metastasis. With a clinical diagnosis of retroperitoneal liposarcoma, we administered preoperative radiation therapy and performed a bilateral tumor resection. On postoperative day 9, a surgical site infection occurred, necessitating incisional drainage and the administration of intravenous antibiotics. The histopathological diagnosis was bilateral myelolipoma of the adrenal glands.

Conclusion: In cases of bilateral retroperitoneal tumors with a large fat component, adrenal myelolipoma should be considered in the differential diagnosis to avoid overtreatment.

Key words: adrenal myelolipoma, case report, differential diagnosis, retroperitoneal liposarcoma, tumor.

Keynote message

This report describes a rare case of giant bilateral adrenal myelolipoma mistaken for retroperitoneal liposarcoma. In cases of bilateral retroperitoneal tumors with a large fat component, the possibility of adrenal myelolipoma should be considered before surgery.

Introduction

Adrenal myelolipomas are uncommon benign adrenal tumors, with 95% of cases being unilateral.[1] The median tumor size is 2–3 cm, and the tumor affects both sexes almost equally.[2] Most patients are asymptomatic, and they do not usually demonstrate tumor growth or adrenal malignancy.

We describe a rare case of giant bilateral adrenal myelolipoma mistaken for retroperitoneal liposarcoma.

Case presentation

A 49-year-old man developed fever, left flank pain, and a large mass on the left abdomen. He had asthma, which was well controlled with inhaled medications and no relevant family history. A plain CT at another hospital showed a 23 × 19 × 13 cm mass with fat-containing components in the retroperitoneal cavity (Fig. 1a,b). The tumor extended from the left side of the diaphragm to the dorsal side of the left kidney, suggestive of a retroperitoneal liposarcoma. A similar 9 × 6 × 4 cm mass was observed in the right adrenal gland, suggestive of adrenal metastasis. MRI showed heterogeneous masses with low signal intensity on T1-weighted images, high signal intensity on T2-weighted images, and internal fat components that were low signal intensity on fat-suppressed T2-weighted images. FDG-PET-CT showed mild heterogeneous uptake (SUV-max 2.4 HU) with clear nodular accumulation

inside the tumor, consistent with malignancy (Fig. 1c). ACTH concentration was 188.0 (normal range: 7.2–63.3) pg/mL, serum cortisol level was 3.07 (normal range: 7.07–19.60) μg/dL, and other blood biochemistry test results were within normal ranges.

With a clinical diagnosis of retroperitoneal liposarcoma, we routinely consider preoperative radiation therapy based on subgroup analysis of liposarcoma in the EORTC-62092 Trial.[3] After discussion with a sarcoma conference, we administered preoperative radiation therapy (total 39.6 Gy: 1.8 Gy × 22 Fraction) to both retroperitoneal cavities and encountered no major adverse events. Thereafter, we performed wide tumor resection comprising a bilateral adrenalectomy, descending colectomy, and distal pancreatectomy using an open approach. The operative time was 647 min, and 1779 mL of blood was lost. Overall, four units of red blood cells were transfused postoperatively. On postoperative day 9, a surgical site infection developed, necessitating incisional drainage and the administration of intravenous antibiotics for 22 days. The patient was discharged on postoperative day 39 with a healthy wound. We prescribed oral hydrocortisone (30 mg daily).

Macroscopically, the largest mass on the left side measured 22 × 13 × 10 cm, and the mass on the right side measured 13 × 8 × 4 cm, with a dark red-to-yellow hemorrhagic tumor (Fig. 2). The histopathological analysis revealed a well-circumscribed mass with a variable mixture of mature adipocytes and large and small erythroid islands, along with myelocytes and megakaryocytes throughout the tumor. No evident atypia was observed in any of the constituent cells. Benign adrenal tissue remained within the both tumors (Fig. 3). No malignancies were observed in the resected tissues. The final diagnosis was myelolipoma of both adrenal glands.

Discussion

To the best of our knowledge, this is the largest bilateral adrenal myelolipoma encountered in Japan. This case suggests that, even in bilateral retroperitoneal tumors with a large fat component, adrenal myelolipoma should be considered in the differential diagnosis to avoid overtreatment. Surgery for retroperitoneal liposarcoma and adrenal myelolipoma differs significantly. Liposarcomas required wide resection, while myelolipoma need only tumorectomy. Especially in this case, we consider the necessity of needle biopsy when it is difficult to make a correct diagnosis.

In most patients with adrenal myelolipoma, the imaging diagnosis is clear, and no further imaging studies are required. However, adrenalectomy is considered if abdominal symptoms or acute hemorrhage is present.[1] Daneshmand *et al*. recommended that symptomatic tumors or myelolipomas over 7 cm should be recommended for surgery because of the spontaneous rupture risk.[4] Primarily because of the

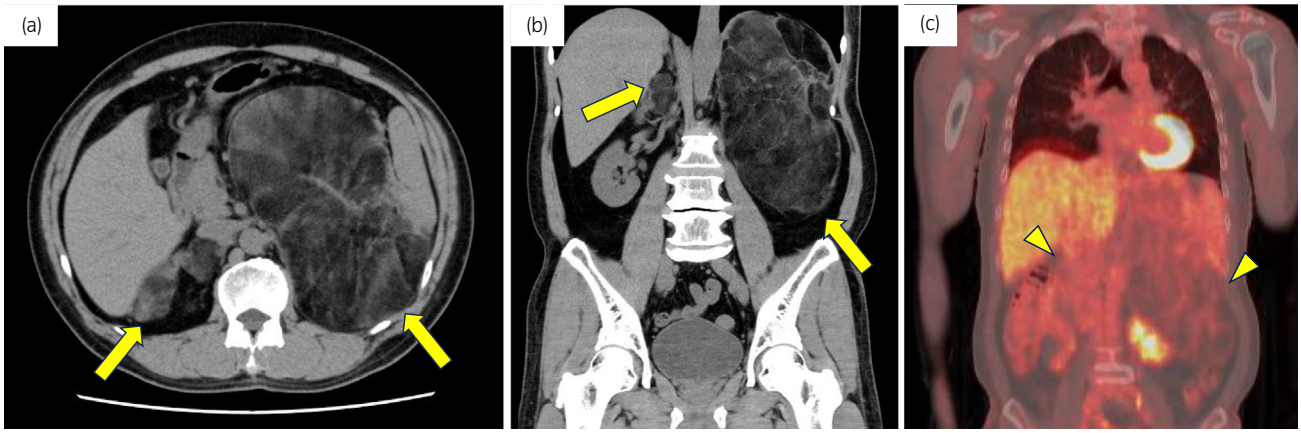

Fig. 1 (a, b) Plain computed tomographic images of the upper abdomen reveal bilateral retroperitoneal masses (yellow arrows). The largest mass on the left side measured 23 × 19 × 13 cm, and the mass on the right side measured 9 × 6 × 4 cm. (c) FDG-PET CT images of the upper abdomen show bilateral retroperitoneal masses (yellow arrowheads) with mild heterogeneous uptake and clear nodular accumulation. The SUV-max value was 2.4 HU.

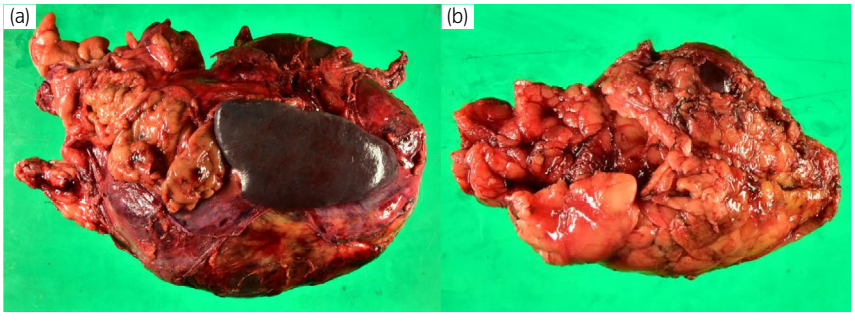

Fig. 2 Macroscopic aspect of the tumor with resected organs. (a) Left adrenal tumor, measuring 22 × 13 × 10 cm, (b) right adrenal tumor, measuring 13 × 8 × 4 cm.

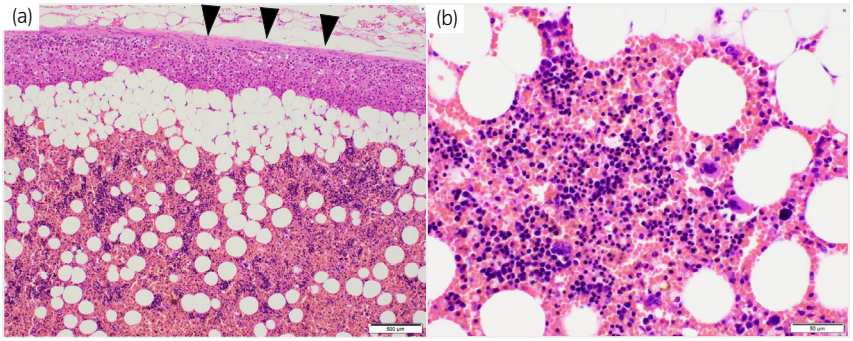

Fig. 3 Histologic appearance of the adrenal myelolipoma (hematoxylin and eosin staining): (a) ×40, (b) ×400. The tumor is composed of mixed adipose tissue and hematopoietic elements with benign adrenal tissue (black arrowheads).

Table 1 Bilateral adrenal myelolipomas larger than 20 cm

	Year	Gender	Age	Larger	Size (cm)	Symptoms	Treatment
1. Kalafatis	1999	Female	34	Right	22 × 18 × 12	Abd. mass	Bilateral adrenalectomy
2. Kalidindi	2006	Male	42	Left	25 × 25 × 14	No	Bilateral adrenalectomy
3. Polamaung	2007	Male	32	Left	27 × 24 × 12	No	Bilateral adrenalectomy
4. Ioannidis	2011	Female	34	Left	24 × 14 × 10	Abd. pain, vomiting	Bilateral adrenalectomy
5. McGeoch	2012	Male	34	Left	23 × 19 × 11	Abd. swelling, discomfort	Bilateral adrenalectomy
6. Al-Bahri	2014	Male	39	Left	25 × 20 × 20	Headache, fatigue, abd. distension	Bilateral adrenalectomy
7. Kale	2015	Male	51	Left	34 × 20 × 13	Back pain, paresthesia	Bilateral adrenalectomy
8. Roger	2021	Male	49	Left	30 × 18 × 13	Abd. pain, vomiting, early satiety	Bilateral adrenalectomy (left cortical sparing)
9. Zhanghuang	2023	Female	45	Left	24 × 18 × 10	Abd. pain, abd. mass	Bilateral adrenalectomy
10. Almutairi	2023	Male	36	Left	39 × 17 × 8	Abd. Pain	Bilateral adrenalectomy
11. Brutvan	2024	Female	42	Right	30 × 25 × 20	Abd. pain, fatigue, nausea	Bilateral adrenalectomy
12. Ito	2024	Male	49	Left	22 × 13 × 10	Abd. pain, fever	Bilateral adrenalectomy

high accuracy of imaging, adrenal biopsy is not necessary in the diagnosis of adrenal myelolipoma.[1]

Twelve cases of giant (>20 cm) and bilateral adrenal myelolipomas have been reported since 1999, including our case (Table 1). Preoperative biopsy was performed only in one case.[5] All previously reported patients underwent a bilateral adrenalectomy because most had large symptomatic tumors and concerns about possible malignancy.[6,7] In some cases, symptomatic adrenal myelolipomas grew in both sides, resulting in bilateral adrenalectomy.[5,8]

The underlying molecular events driving adrenal myelolipoma are poorly understood.[1] Decmann *et al*. reported that 10% of patients with adrenal myelolipoma had congenital adrenal hyperplasia.[9] Adrenal myelolipoma is usually hormonally inactive and most patients with myelolipoma have normal levels of ACTH. However, some previous studies suggested that ACTH influences the development of myelolipomas, and reported overexpression of ACTH and androgen receptor immunoreactivity in the tumor.[10,11] In this case, the ACTH level was high and serum cortisol level was low. This may be related to the bilateral tumor growth.

Adrenal myelolipoma is diagnosed using imaging, either CT or MRI with or without contrast enhancement, by radiological identification of the fatty components. CT attenuation is very low, and it appears dark on CT and white on T1-weighted and T2-weighted MRI scans. As the myeloid components increase, the attenuation of the CT increases and many myelolipomas measure between −50 and −20 HU.[1] In this case, the tumors were bilateral, huge, multilobulated, heterogeneous, and measured between −100 and 60 HU, which is the main reason for the misdiagnosis of retroperitoneal liposarcoma. Occasionally, the imaging appearance of myelolipomas is similar to that of liposarcomas. Consequently, myelolipomas are often misdiagnosed as liposarcomas on imaging studies.[12,13]

Shimoda *et al*. suggested the usefulness of FDG-PET in differentiating between myelolipoma and liposarcoma.[12] In a report of 10 patients with myelolipoma, the mean maximum standardized uptake (SUV-max) values were 0.7 HU in the fat component and 1.4 HU in soft tissue myeloid component.[14] Meanwhile, in the report of 136 patients with retroperitoneal sarcoma, SUV-max was different between the histologic subtype, and median SUV-max for well-differentiated liposarcoma, dedifferentiated liposarcoma, and leiomyosarcoma were 2.32, 6.32, and 12.04 HU, respectively.[15] In the present case, the SUV-max value was 2.4 HU, which was similar to well-differentiated liposarcoma in the previous report. FDG-PET might be useful in identifying myelolipoma and dedifferentiated liposarcoma. If the diagnosis of adrenal myelolipoma cannot be determined, needle biopsy is considered to avoid overtreatment.[3]

Conclusions

We reported the case of a male patient with giant bilateral adrenal myelolipoma that was mistaken for a retroperitoneal liposarcoma. Our case highlights the importance of considering the possibility of adrenal myelolipoma before surgery

when bilateral retroperitoneal tumors with a large fat component are observed.

Acknowledgments

None.

Author contributions

Kana Ito: Investigation; methodology; project administration; writing – original draft. Ryo Yamashita: Supervision; writing – review and editing. Yuma Sakura: Writing – review and editing. Hideo Shinsaka: Writing – review and editing. Masafumi Nakamura: Writing – review and editing. Masato Matsuzaki: Writing – review and editing. Masashi Niwakawa: Writing – review and editing. Tsubasa Hiraki: Writing – review and editing.

Conflict of interest

The authors declare no conflict of interest.

Informed consent

Written informed consent was obtained from the patient.

Approval of the research protocol by an Institutional Review Board

Not applicable.

Registry and the Registration No. of the study

Not applicable.

Animal studies

Not applicable.

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