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A case of perirenal extra-adrenal myelolipoma mimicking liposarcoma

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ABSTRACT

Myelolipoma is a benign tumor composed of mature adipose tissue and normal hematopoietic components. It usually occurs in the adrenal glands but rarely in the extra-adrenal region. However, it is difficult to differentiate extra-adrenal myelolipoma from well-differentiated liposarcoma on the basis of the radiological findings. We report the case of a 66-year-old male with perirenal and extra-adrenal myelolipoma who underwent radical tumor resection with nephrectomy after a preoperative diagnosis of liposarcoma. Intraoperative assessment by the surgeon and intraoperative pathological evaluation are important considering the divergent prognoses of myelolipoma and liposarcoma.

1. Introduction

Myelolipoma is one of the benign tumors occurring in the adrenal glands. It is composed of mature adipose tissue and normal hematopoietic components that produce trilineage cells including megakaryocytes, erythroblasts, and myeloblasts. Although myelolipoma typically occurs in the adrenal glands, rarely, it may be found in the extra-adrenal regions, and is then referred to as extra-adrenal myelolipoma. In approximately 50% of cases, extra-adrenal myelolipoma is localized in the presacral region, but rarely occur in perirenal retroperitoneum.[1] When extra-adrenal myelolipoma occurs in the perirenal retroperitoneum, radiological differentiation from retroperitoneal liposarcoma is quite challenging. We report the case of a 66-year-old male who underwent radical surgery for a retroperitoneal tumor suspected to be a liposarcoma, which turned out to be an extra-adrenal myelolipoma. Considering the divergent prognosis of myelolipoma and liposarcoma, intraoperative assessment by the surgeon and intraoperative pathological evaluation are important.

2. Case presentation

A 66-year-old asymptomatic male patient was referred to our hospital because of an incidentally discovered retroperitoneal tumor. The patient had a history of alcoholic liver injury, spinal disc herniation, diabetes mellitus, hypertension, and dyslipidemia. He was a current smoker, with a Brinkman index of 500. Laboratory tests showed elevated levels of hemoglobin (18.7 g/dL), hemoglobin A1c (7.7%), aspartate aminotransferase (AST; 74 U/L), alanine transaminase (ALT; 132 U/L), gamma–glutamyl transpeptidase (GTP; 209 U/L), and C-reactive protein (0.21 mg/dL).

Computed tomography revealed a heterogeneously enhanced tumor with a diameter of 8 cm in the retroperitoneum, on the caudal side of the left kidney. The tumor consisted of a high-density solid and macroscopic fat components (Fig. 1A). Fluorodeoxyglucose positron emission tomography (FDG-PET) revealed slight FDG uptake in the retroperitoneal tumor (maximum standardized uptake value [SUVmax], 1.8) (Fig. 1B). The tumor showed heterogeneously high signal intensity on T2-weighted magnetic resonance imaging (Fig. 2A), which was completely suppressed on fat-suppressed T2-weighted images (Fig. 2B). The tumor showed a weak signal on diffusion-weighted imaging (Fig. 2C). Based on these radiological findings, the initial differential diagnosis was a retroperitoneal dedifferentiated liposarcoma.

Based on the large tumor size and the possibility of liposarcoma, open transabdominal excision of the retroperitoneal tumor was performed. As the tumor was tightly adhered to the perinephric fat and involved the left ureter, left nephrectomy was performed simultaneously. On gross examination, the tumor measured 10 × 10 × 3.5 cm and was composed of yellow fat tissue mixed with reddish-brown components. Histologically, the tumor was composed of mature adipose and myeloid tissue with trilineage hematopoiesis (Fig. 3A and B).

Abbreviations: ACTH, adrenocorticotropic hormone; ALT, alanine transaminase; AST, aspartate aminotransferase; FDG-PET, fluorodeoxyglucose positron emission tomography; MDM2, murine double minute 2 protein; GTP, gamma-glutamyl transpeptidase; SUVmax, maximum standardized uptake value.

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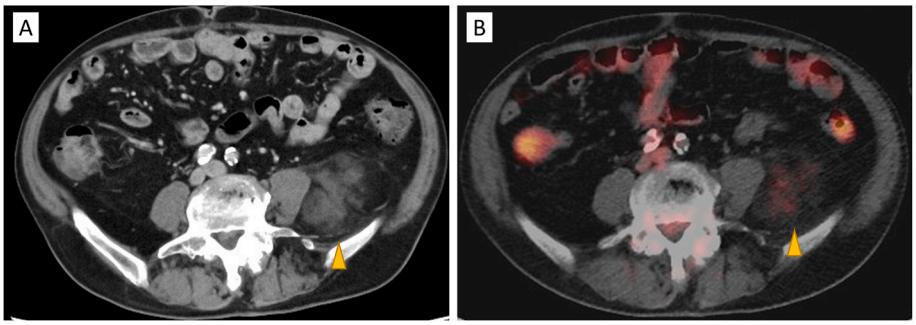

Fig. 1. Computed tomography (A) and fluorodeoxyglucose (FDG) positron emission tomography (B) revealing a heterogeneous tumor with a macroscopic fat component and slight FDG uptake.

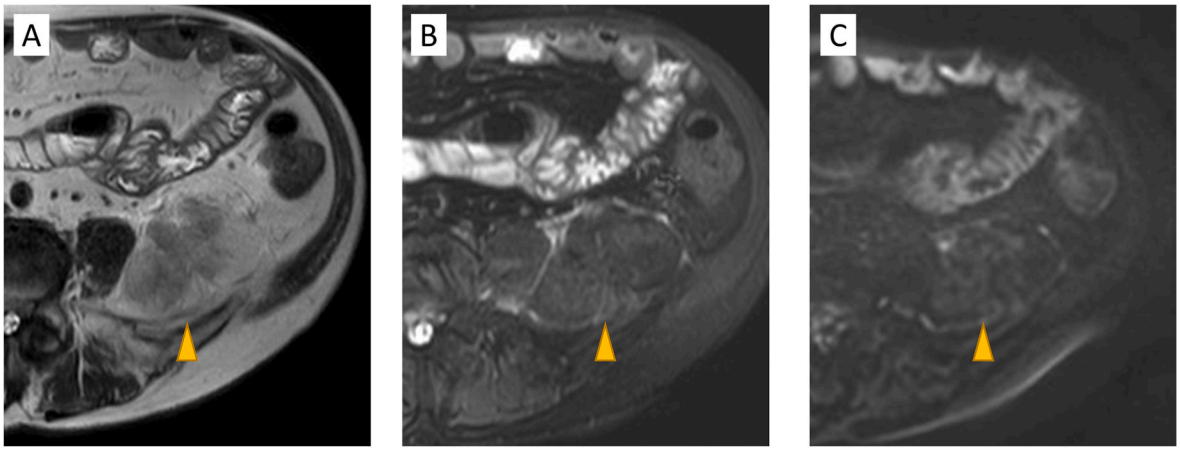

Fig. 2. T2-weighted (A), fat suppressed T2-weighted (B), and diffusion-weighted (C) magnetic resonance imaging reveal that the tumor is composed of fat component with relatively low cellular density.

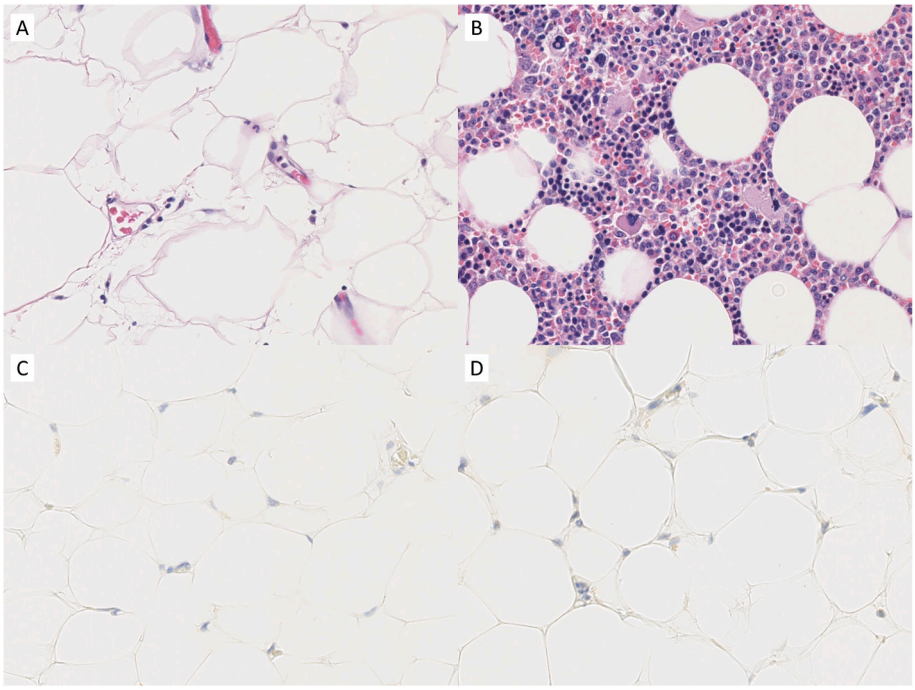

Fig. 3. Pathological and immunohistochemical findings of the tumor. The tumor is composed of mature adipose tissue (A) and myeloid components (B). The tumor is negative for cyclin-dependent kinase 4 (CDK-4) (C) and murine double minute 2 protein (MDM-2) (D).

Immunohistochemical analysis revealed negative staining for cyclin-dependent kinase 4 (CDK-4) and murine double minute 2 protein (MDM-2) (Fig. 3C and D), suggesting that a liposarcoma was unlikely. Based on these findings, the definitive pathological diagnosis was extra-adrenal myelolipoma. The patient has been doing well without recurrence for one year after surgery.

3. Discussion

Myelolipoma is a rare benign tumor that mostly occurs in the adrenal gland. Extra-adrenal myelolipoma is extremely rare, with fewer than 100 reported cases. Although extra-adrenal myelolipoma may involve various regions, including the perirenal retroperitoneum as in this case, as well as the mediastinum, lung, liver, spleen, bone, and kidney, approximately 50% of extra-adrenal myelolipoma occurs in the presacral region.[1] Therefore, macroscopic fat-containing tumors in the adrenal gland or presacral space can be easily diagnosed as benign myelolipoma. However, the radiological differential diagnosis of extra-adrenal myelolipoma and liposarcoma may be quite challenging when these tumors develop in other lesions. Preoperative pathological diagnosis is beneficial because it largely affects the treatment strategy. However, the risk of tumor seeding in biopsy tracts should be considered, and a definitive diagnosis can sometimes be difficult with needle biopsy if the biopsy specimens include only fat tissue without myeloid components. In fact, there have been several case reports of extra-adrenal myelolipoma in which a definitive preoperative diagnosis could not be made.[2]

In the current case, the tumor showed slight FDG uptake, with an SUVmax of 1.8. In a report of 10 patients with myelolipoma,[3] the average SUV-max was 0.7 in the fat-component and 1.4 in soft tissue myeloid component. In contrast, in 20 patients with liposarcoma,[4] the average SUV-max was 3.15 in well-differentiated liposarcoma and 9.23 in dedifferentiated liposarcoma. Although further case studies are required, FDG-PET may be useful in differentiating between myelolipoma and liposarcoma.

The etiology of myelolipoma is unclear owing to their rarity. A possible association between erythropoietin and the development of myelolipoma has been suggested,[5] as elevated erythropoietin levels in patients with chronic anemia may stimulate the development of adrenal myelolipoma. An association between elevated adrenocorticotropic hormone (ACTH) concentration and an increased risk of adrenal myelolipoma has also been suggested,[5] as these lesions are frequently reported in patients with Cushing disease or congenital adrenal hyperplasia. The current patient did not have anemia or symptoms of Cushing disease; thus, the etiology could not be determined.

Classically, asymptomatic and small (<4 cm in diameter) myelolipoma is managed conservatively with routine radiological surveillance, and surgical resection is reserved only for symptomatic or large tumors. However, extensive surgery is required for liposarcoma, including the well-differentiated, because of its worse prognosis. Whether the differential diagnosis is either myelolipoma or liposarcoma affects the treatment strategy; therefore, preoperative pathological diagnosis with needle biopsy is important. In cases not definitively diagnosed by preoperative needle biopsy, intraoperative assessment by the surgeon and intraoperative pathological evaluation are important because the surgical strategies for myelolipoma and liposarcoma vary widely.

4. Conclusion

We report a case of extra-adrenal myelolipoma that occurred in the perirenal region. Extra-adrenal myelolipoma that occur in the retroperitoneal region is sometimes difficult to differentiate from liposarcoma; thus, intraoperative assessment by the surgeon and intraoperative pathological evaluation are important.

Funding

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Declaration of interest

None.

Informed consent

Written informed consent was obtained from the patient for the publication of this case report and accompanying images.

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