Extrarenal retroperitoneal angiomyolipomas (AML) are extremely rare, therefore they may present a diagnostic challenge. In this paper, the authors describe a case of a huge retroperitoneal AML in a 49-year-old woman who presented with sudden abdominal pain. Computed tomography revealed the presence of a large, round, fatty mass in the retroperitoneal space, which was easily removed by surgery. The mass was well encapsulated and dark yellow on the cut surface. Microscopically, the tumor was exclusively composed of adipose tissue with frequent multivacuolated, lipoblast-like cells masquerading as well differentiated liposarcoma. In addition, there were many clear, epithelioid cells present, especially around the small blood vessels, which were reactive for HMB-45 and smooth muscle actin.

**Key Words**: Angiomyolipoma; Liposarcoma; Retroperitoneal neoplasms

### CASE REPORT

A 49-year-old woman visited a local clinic due to a sudden onset of abdominal clamping pain that had not responded to oral treatment after several days. The local clinic conducted CT, which revealed the presence of a huge abdominal mass, therefore, the woman was admitted to our clinic for evaluation of the mass. Abdominal CT showed a well-delineated large intraabdominal fatty mass that contained multiple tortuous vascular structures inside the mass (Fig. 1A). The mass, which appeared to originate from the left upper quadrant of the retroperitoneum, had pushed the stomach, pancreas, and spleen aside. Laparotomy revealed a huge, round, well encapsulated mass that was 16 × 15 × 8 cm and had a thick fibrous capsule with a glistening outer surface, as well as several dilated vessels. The cut surface of the mass was soft and dark yellow in color and showed hemorrhagic areas (Fig. 1B). Microscopically, the tumor was exclusively composed of variably sized adipocytes (Fig. 1C) with frequent mono- or multi-vacuolated cells resembling lipoblasts, though there was no evidence of nuclear atypia or pleomorphism. Additionally, although focal areas of smooth muscle proliferation were observed, the large, tumorous blood vessels that were seen on a previous CT scan were not found, despite extensive sampling. It is also important to note that there were single or small clusters of epithelioid cells observed around small blood vessels and between fat cells (Fig. 1D). These epithelioid cells had ovoid vesicular nuclei with clear cytoplasm, some of which contained eosinophilic cytoplasmic granules and were strongly reactive for smooth muscle actin (SMA; 1:100, Dako) and HMB-45 (1:40, Dako) (Inset in Fig. 1D). Based on these histologic and immunohistochemical findings, a final diagnosis of lipomatous AML was made. The patient's postoperative course was uneventful and she...
was discharged 5 days postoperatively.

**DISCUSSION**

AML is an uncommon mesenchymal neoplasm that is classically composed of three tissue components; convoluted thick-walled blood vessels, smooth muscle cells, and mature adipose tissue. Although the proportions of these tissue types are quite variable, if one particular component predominates, it may be subdivided into angiomatous, myomatous, or lipomatous type tissue. Although most AMLs arise in the kidney, they can also arise in the liver, nasal cavity, oral cavity, heart, colon, lung, and skin. However, a retroperitoneal location is unusual, with only approximately 10 cases being reported in the English literature.\(^2\)\(^-\)\(^5\)

In this case, the main differential diagnosis was initially liposarcoma based on the radiologic and pathologic findings. This initial diagnosis was made because liposarcoma is the most common lipogenic tumor of the retroperitoneum, and the tumor was exclusively composed of adipose tissue that contained many multivacuolated, lipoblast-like cells. However, hyperchromatic, atypical stromal cells, which are typical of well-differentiated liposarcoma, were not observed. Additionally, in contrast to liposarcoma, small clusters of clear, epithelioid cells (so called perivascular epithelioid cells, PECs) were frequently identified around the blood vessels. These epithelioid cells were found to be immunoreactive for HMB-45 and SMA, which aided in the differentiation of this case from liposarcoma. Although this case may have also been diagnosed as myolipoma, only limited areas of smooth muscle proliferation were observed, and the tumor in this case contained many HMB-45 positive epithelioid cells, which are not found in myolipoma.

Previously described retroperitoneal AMLs have been huge and shown fat signals on the CT scans, therefore they have com-
monly been diagnosed as liposarcoma. In addition, most previ-
ously described retroperitoneal AMLs have exhibited histologi-
cal features typical of AML, and have possessed all three of the
aforementioned tumor components, therefore the histological
diagnoses in these cases were straightforward. In contrast, mono-
typic lipomatous AML, as was observed in the present case, has
only been reported once, and may present a diagnostic chal-
lenge, even to experienced pathologists. Most retroperitoneal
AMLs appear to be lipogenic on CT scans, and are difficult to
differentiate from liposarcoma. However, a tortuous hypervas-
cularity is a common feature of retroperitoneal AMLs, regard-
less of histology. Additionally, Wang et al. previously reported
that CT characteristics, including linear vascularity, aneurysmal
dilatation, bridging vessels, and beak sign, are helpful for diag-
nosing retroperitoneal AMLs.

Retroperitoneal AMLs are benign, therefore complete surgi-
cal excision is considered sufficient to cure this disease. In addi-
tion, because these tumors have a propensity to bleed, preopera-
tive selective arterial embolization can be helpful for reducing
intraoperative bleeding. Moreover, although most tumors require
surgical excision, one exceptional case was successfully treated
by simply embolizing the primary feeding vessels.

In conclusion, AML rarely occurs in the retroperitoneum, and
it may be difficult to differentiate AML from liposarcoma, espe-
cially when the tumor is histologically of the monotypic lipo-
matous type. However, intratumoral hypervascularity may be
an important radiological feature that favors a diagnosis of AML.

REFERENCES

1. Weiss SW, Goldblum JR. Enzinger and Weiss’s soft tissue tumor.
2. Hruban RH, Bhagavan BS, Epstein JI. Massive retroperitoneal angio-
myolipoma: a lesion that may be confused with well-differentiated
3. Shimada S, Harada H, Ishizawa K, Hirose T. Retroperitoneal lipo-
matous angiomyolipoma associated with amyloid deposition mas-
querading as well-differentiated liposarcoma. Pathol Int 2006; 56:
638-41.
4. Tseng CA, Pan YS, Su YC, Wu DC, Jan CM, Wang WM. Extrarenal
retroperitoneal angiomyolipoma: case report and review of the lit-
5. Neary P, Mathews R, Harries R, McDonald A, Monson JR. Extrare-
nal retroperitoneal angiomyolipoma: management of a rare benign
characteristics that differentiate angiomyolipomas from liposarco-
7. Murphy DP, Glazier DB, Chenven ES, Principato R, Diamond SM.
Extrarenal retroperitoneal angiomyolipoma: nonoperative manage-