

RADIOGRAPHIC QUIZ

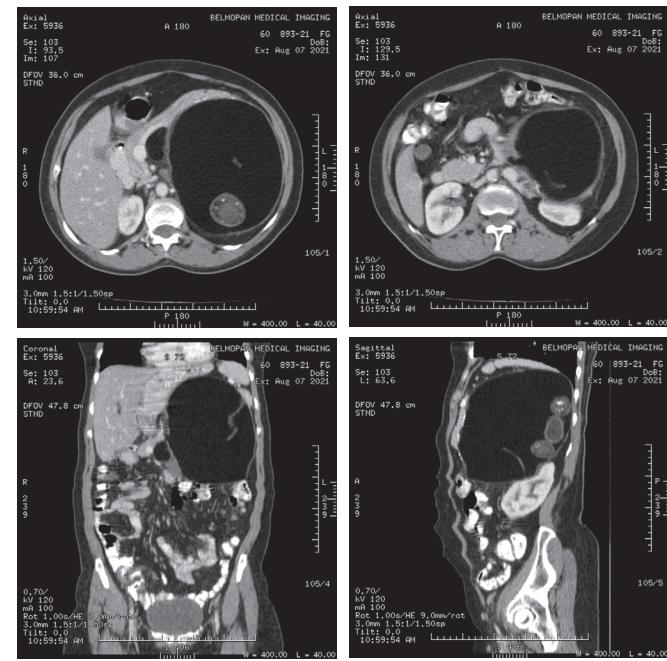
From page 16

■ BRIEF OVERVIEW OF THE DISEASE.

Angiomyolipoma (AML) is a benign tumor primarily containing fat, atypical blood vessels and smooth muscles in varying proportions. The common site of AML is kidney and it presents primarily as an intra-renal mass. Occasionally, a renal AML may outgrow exophytically in the retroperitoneum and closely mimics a liposarcoma due to the high fat content on a radiological imaging. The diagnostic dilemma is further compounded as the core-cut biopsy may also be erroneously reported as liposarcoma. This may result in an aggressive surgery, and at times multi-visceral resections, to achieve a margin-negative resection. A post-resection histopathological diagnosis of AML may come as a surprise for both the surgeons and the patients, and may even lead to litigations.

Liposarcoma and exophytic renal angiomyolipoma represent two retro-peritoneal masses that contain fatty elements. On occasion, their appearances may be so similar that they can be confused on imaging and even sometimes at histologic examination. However, their differentiation is important because the prognosis and treatment are different. For liposarcomas, surgical resection—usually with the adjacent kidney—is necessary, although complete surgical removal is difficult, and recurrence is common. Angiomyolipomas, although benign, may hemorrhage and require emergent treatment (embolization or surgery) if life-threatening bleeding occurs, but they do not necessarily require surgery. Liposarcomas, which occur slightly more frequently in men than in women, are among the most common primary retroperitoneal malignancies, with the perinephric region a frequent location. Angiomyolipoma (renal hamartoma), a benign tumor, is more common in women. These tumors may grow to be large and bulky, extending into the perinephric space.

In a retrospective study of CT images of 15 large exophytic renal AMLs and 12 well-differentiated perirenal liposarcomas, Israel et al suggested that a defect in the renal parenchyma combined with the presence of the enlarged vessels may help differentiate an exophytic AML from a retroperitoneal sarcoma. In a study of 11 patients with the perirenal liposarcoma and 9 patients with the giant exophytic AMLs, Ellingson et al also concluded that the presence of tumoral vessel extending into the renal cortex or a renal parenchyma defect at the site of the tumor contact points strongly towards the diagnosis of AML while intra-tumoral calcification favors the presence of liposarcoma. However, Wang et al suggested that the intra-tumoral calcification can be seen in both AML and liposarcoma. They highlighted that the characteristics of a perinephric AML are intratumorally linear vascularity, aneurysmal dilatation, bridging vessel sign, hematoma, beak sign, and discrete intra-renal fatty tumors while the CT characteristics of a perinephric liposarcoma are a non-fatty soft tissue mass. Other close differentials of an exophytic AML



include lipoma, leiomyoma with fatty change or a fat containing renal cell carcinoma.

Because renal angiomyolipomas may grow exophytically, most of the neoplasm may extend into the perirenal space. However, because these angiomyolipomas arise from the kidney, a defect will be present in the renal parenchyma at their origin. In comparison, liposarcomas arise in the retroperitoneal fat, including the perirenal fat within Gerota's fascia. They are often closely associated with the renal capsule and are sometimes called capsular liposarcomas. When these lesions grow, they displace, compress, and distort the kidney but usually do not invade the adjacent renal parenchyma. Therefore, liposarcomas do not cause a defect in the renal parenchyma, and the interface of the lesion with the kidney is smooth.

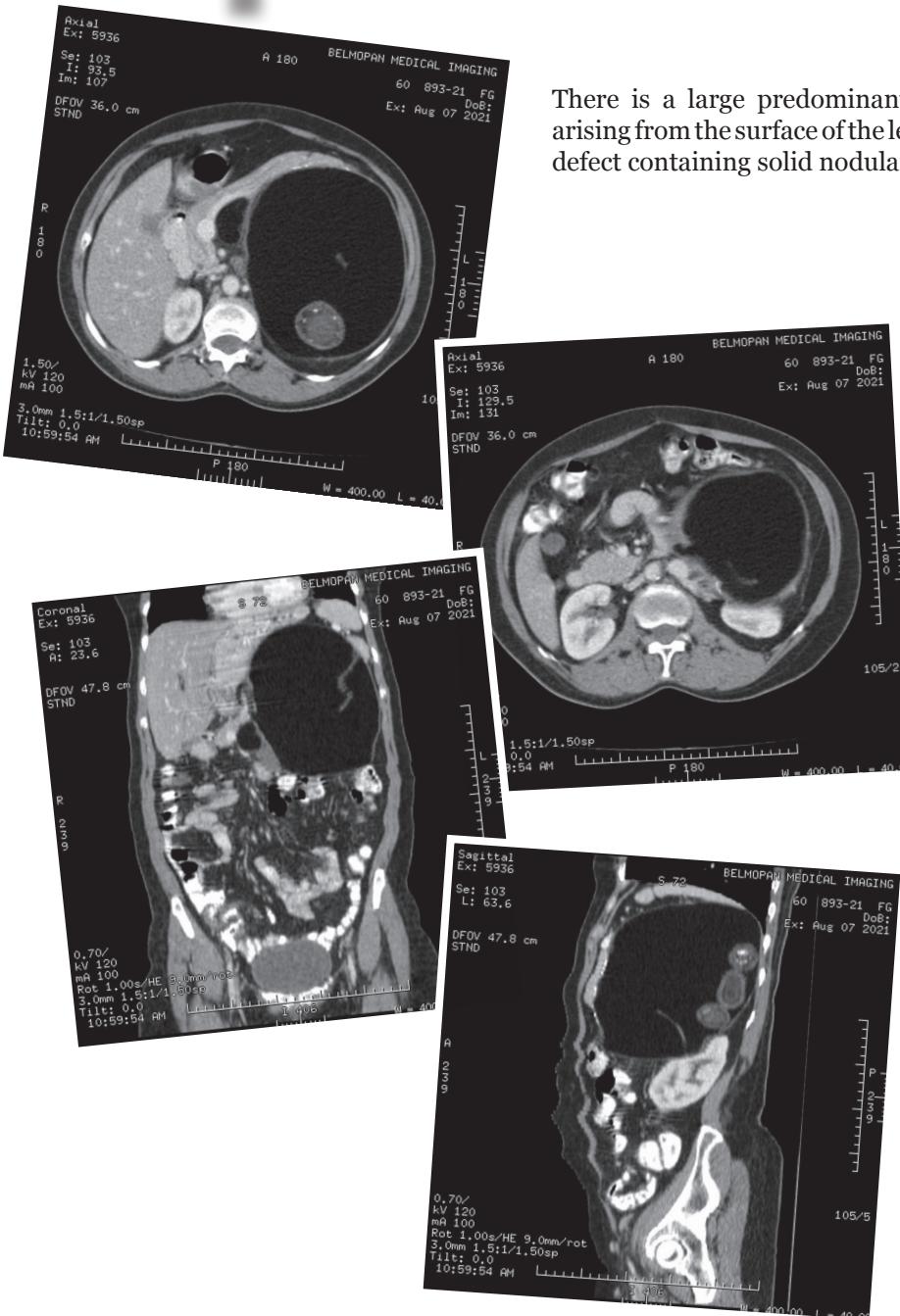
We conclude that a giant exophytic renal AML can pose a serious diagnostic challenge and may be confused with a retroperitoneal liposarcoma. A discordance of the radiological and core biopsy findings in a suspected case of exophytic renal AML must lead to re-evaluation of the case and repeat biopsies may further clarify the diagnosis.

■ REFERENCES:

World J Clin Oncol. 2018 Nov 10; 9(7): 162–166. Published online 2018 Nov 10. doi: 10.5306/wjco.v9.i7.162

CT Differentiation of Large Exophytic Renal Angiomyolipomas and Perirenal Liposarcomas. Gary M. Israel¹, Morton A. Bosniak, Chrystia M. Slywotzky and Robert J. Rosen. September 2002, Volume 179, Number 3

Answer to Radiographic Quiz



There is a large predominantly fat containing left retroperitoneal mass arising from the surface of the left kidney causing a smooth renal parenchyma defect containing solid nodular components and intratumorally vessels.

What is the main abnormality?

- Solid nodules with calcifications in the left hypochondrium
 Large left retroperitoneal predominantly fatty mass arising from the left kidney
 Large left renal cystic mass

There are inflammatory changes in the peritumoral fat

- True
 False

There are vessels within the tumoral mass

- True
 False

The tomographic appearance suggests a giant exophytic left renal angiomyolipoma or a retroperitoneal liposarcoma.

- True
 False

The presurgical differentiation between giant exophytic renal angiomyolipoma and retroperitoneal sarcoma is considered an important point.

- True
 False

Sharp defect in renal parenchyma and enlarged vessels within the mass favor the diagnosis of renal angiomyolipoma.

- True
 False