LARGE EXOPHYTIC RENAL ANGIOMYOLIPOMA

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ABSTRACT

A case of large exophytic renal angiomyolipoma in a 24-year-old female patient is presented. Abdominal CT scans showed the characteristic findings: a large exophytic well-demarcated fat density mass with sharp defect in renal parenchyma, enlarged intratumoral blood vessels and presence of additional intrarenal angiomyolipomas.CT is an accurate and clinically useful method to evaluate renal angiomyolipoma and provide the differential diagnostic information.

INTRODUCTION

Angiomyolipoma is a benign renal neoplasm that consist of varying amounts of mature adipose tissue, smooth muscles and thick-walled blood vessels. This tumor may grow to be large and bulky, extending into the perirenal space and represent retroperitoneal mass that contain fatty element. A large predominately exophytic angiomyolipoma may be difficult to be distinguished from a well-differentiated perirenal liposarcoma because both are large fat -containing lesions and their CT appearances may be so similar. Their differentiation are important because the prognosis and treatment are different.[1]

A case of large exophytic renal angiomyolipoma with the imaging findings and CT characteristics that lead to accurate distinction from retroperitoneal perirenal liposarcoma is presented.

CASE REPORT

A 24-year-old female patient who had a history of exploratary laparotomy due to rupture of ectopic pregnancy about 1 month ago, presented with left upper quadrant pain for 2 months. Physical examination revealed large abdominal mass size about 10x20 cm. at left upper abdomen with mild tenderness. The rest of the physical examination were normal. Routine laboratory values were within normal limits.

An abdominal CT study was performed. It demonstrated a large well demarcated homogeneously fatty tumor in left retroperitoneum, measuring 25x18x10 cm., surrounding and displacing left kidney upward and anteriorly with sharp defect in the lower pole of left kidney. Multiple small fatty tumors, size about 0.5-1 cm. at the lower pole of left kidney are also seen.(Fig.1) Post contrast study reveals enhancement of intratumoral enlarged vessels. (Fig. 2-3) The CT findings are suggestive of a large predominately exophytic renal angiomyolipoma at the lower pole of left kidney with additional multiple small renal angiomyolipoma in the ipsilateral kidney.

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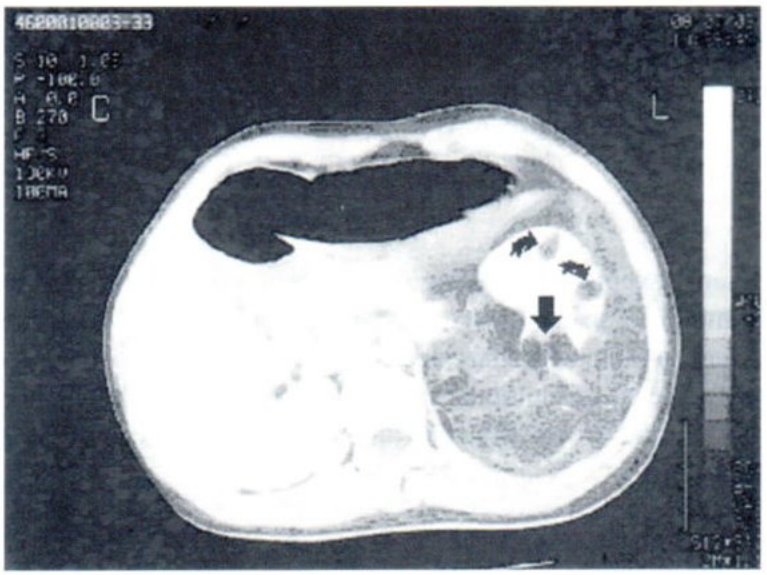

Fig.1 Contrast-enhanced CT scan of the abdomen shows a large well demarcated fatty tumor in the left retroperitoneum, surrounding and displacing left kidney upward and anteriorly with sharp defects in the renal parenchyma (arrow) and additional multiple small fatty tumors at lower pole of left kidney.(curve arrows)

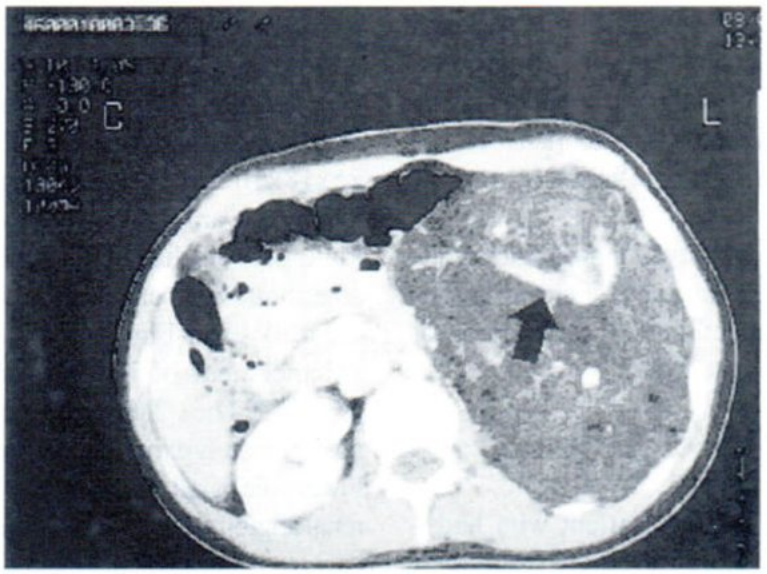

Fig.2 Contrast-enhanced CT scan presented inferior to the left kidney shows associated with enlarged vessels. (arrow)

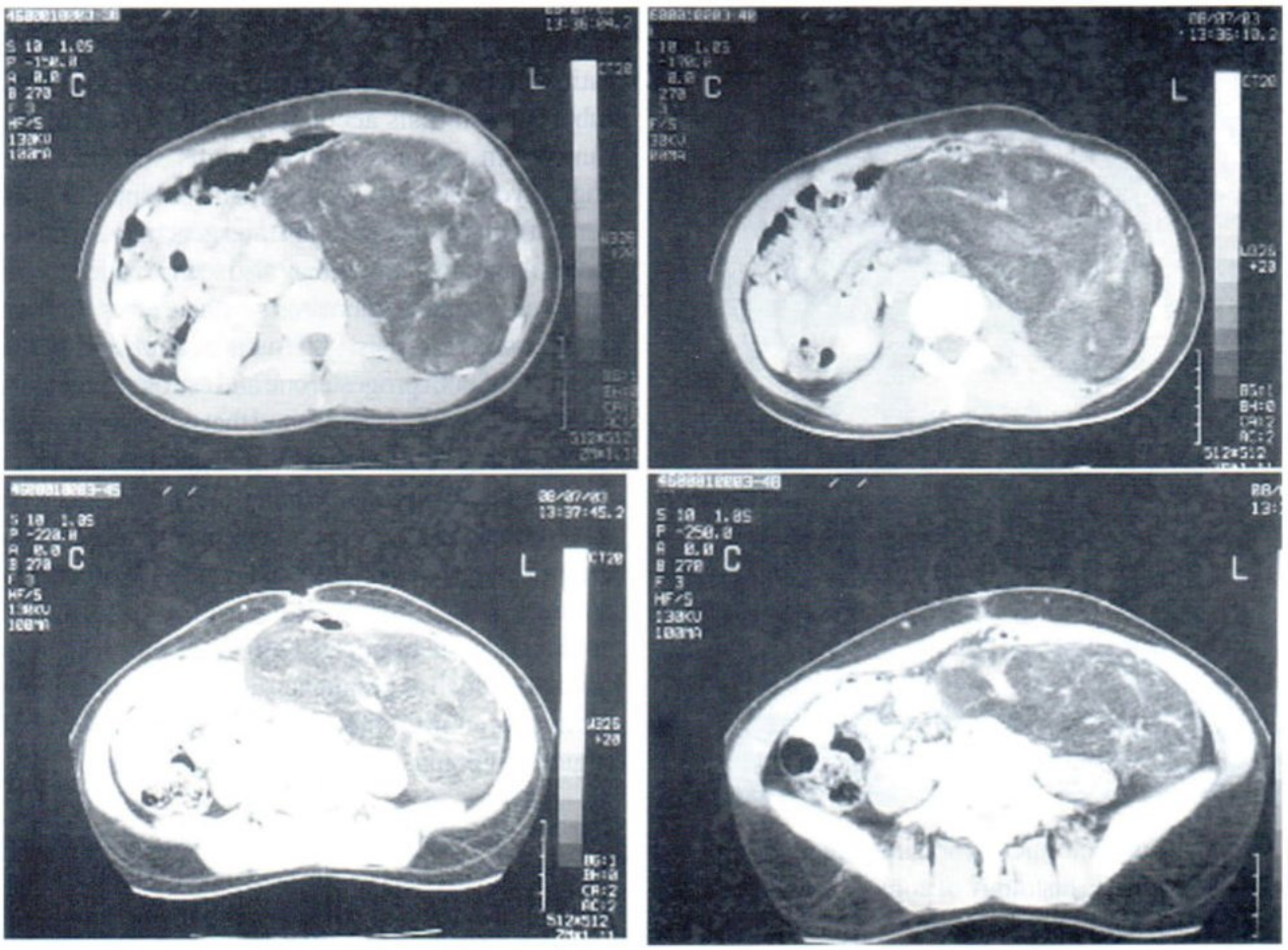

Fig.3 Contrast-enhanced CT scan obtained inferior to Figure 2 shows further extension of the lesion into the pelvic cavity.

On exploratory laparotomy, a large well -capsulated yellow-whitish mass size about 30x20x10 cm. and weighed 2300 g. was found arising from the lower pole of left kidney and located in the left retroperitoneal space. The tumor was removed completely.

The histopathologic study revealed the triphasic appearance, with smooth muscle,adipose tissue and thick-walled blood vessels.(Fig.4) The histopathologic diagnosis was angiomyolipoma.

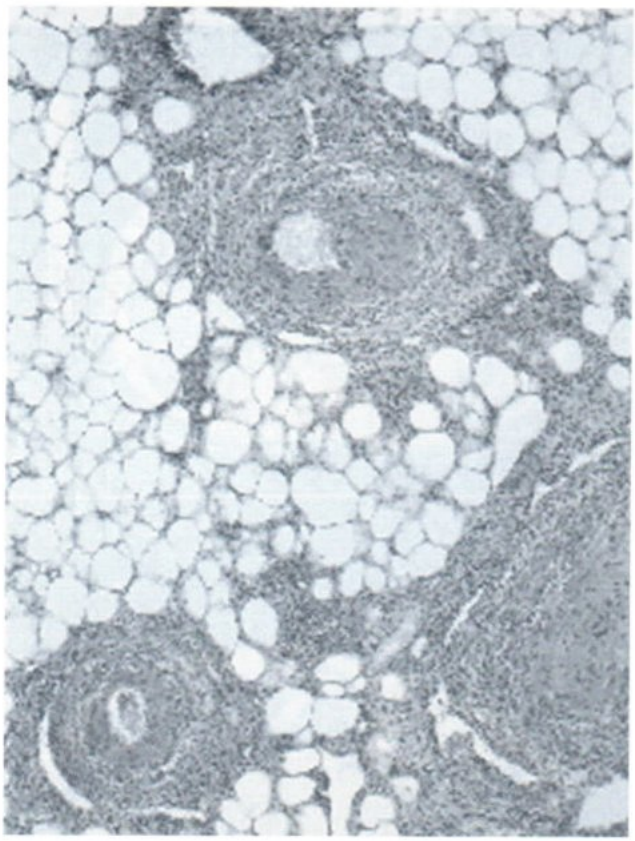

Fig.4 This photomicrograph illustrated the typical triphasic histology of angiomyolipoma, with smooth muscle, adipose tissue and thick walled blood vessels.

DISCUSSION

The first histologic description of an angiomyolipoma appeared in the literature in 1911 reported by Fischer.[2] The term hamartoma (a benign mass composed of disorganized tissues normally found in an organ) is sometimes used, but choriostoma (a benign mass composed of disorganized tissues not normally found in the organ) is a more appropriate term because smooth muscle is not normally found in renal parenchyma.[3] Two types of angiomyolipoma are described: isolated angiomyolipoma and angiomyolipoma associated with tuberous sclerosis. Isolated angiomyolipoma occurs sporadically and accounts for 80 percent of the tumors.[4] In symptomatic patient without tuberous sclerosis, the tumors are usually large, solitary lesions occurring in women between the ages of 30 and 60 years, with a female to male ratio of 2.6 to 1. Angiomyolipoma associated with tuberous sclerosis accounts for 20 percent of the tumors. In patients with tuberous sclerosis, the angiomyolipoma are usually small, multiple (13 to 30 percent), bilateral (15 percent) and generally asymptomatic. Angiomyolipoma may also seen in patients with pulmonary lymphangiomatosis without other stigmata of tuberous sclerosis.[5] L'Hostis et al observed the presence of both progesterone and estrogen receptors in angiomyolipomas and found that progesterone and estrogen immunoreactive angiomyolipomas were predominantly found in women and in patients with tuberous sclerosis. These findings may further explain the more aggressive nature of the disease process in patients with tuberous sclerosis, the hormonal potentiation of tumor growth and hemorrhage in conditions such as pregnancy and the overwhelming female predominance in the sporadic form of angiomyolipoma without tuberous sclerosis.[7]

Angiomyolipoma is a round or oval tumor that arise in both the renal cortex and medulla and elevates the renal capsule. It grows by expansion and local invasion. In about 25 percent, the tumor has a predominantly exrarenal growth pattern that extends to or even through the renal capsule into the perirenal compartment.[2] It is considered benign, but rare cases extension into renal vein and/or inferior vena cava and deposits in the renal lymphnodes are reported. Most small lesions are asymptomatic and incidental findings on images are detected. As many as 40 percent are symptomatic, manifestation with symptoms of abdominal and flank pain, nausea, vomiting and fever may be detected. Common findings described include a palpable mass, abdominal tenderness, hematuria, anemia, shock, hypertension, UTI and renal failure. These signs and symptoms are usually a result of mass effect and hemorrhage.

Accurate diagnosis of angiomyolipoma can be made in virtually all cases using CT, which is very sensitive to the low attenuation of fatty tissue within

the tumor, typically less than -20 HU at nonenhanced CT.[8] Angiomyolipoma typically join the normal renal tissue at an acute angle, rarely obstruct the calyceal system, and range in size from less than 2 cm. to more than 8 cm., when detected. After contrast medium injection, portions of tumor may be enhanced, but fatty tissue do not increase in density. Angiomyolipoma commonly contains enlarged vessels that can be seen on contrast-enhanced CT.

Large exophytic renal angiomyolipoma represents a retroperitoneal mass that contain fatty element, with CT appearances similar to retroperitoneal liposarcoma. Liposarcoma which occur slightly more frequently in men than in women, are among the most common primary retroperitoneal malignancies, with the perirenal region as a frequent location. Their differentiation is important because the prognosis and treatment are different. Gary M. Israel et.al retrospectively analyzed CT images of exophytic renal angiomyolipomas and well-differentiated retroperitoneal liposarcoma and described the important imaging findings; sharp defect in the renal parenchyma, the presence of enlarged vessels and associated angiomyolipomas that enable accurate differentiation angiomyolipoma from retroperitoneal perirenal liposarcoma.[1]

Renal angiomyolipoma arise from the kidney, a defect will be present in the renal parenchyma at its origin and commonly contains enlarged vessel. Liposarcoma do not cause a defect in renal parenchyma and the interface of the lesion with the kidney is smooth.

Well-differentiated liposarcoma is relatively avascular, and its vessels are not usually enlarged. The presence of enlarged vessels in renal angiomyolipoma is not as important in diagnosis as the presence of a defect in renal parenchyma but it is a significant ancillary finding. Renal angiomyolipoma may be multiple, even without associated tuberous sclerosis. The presence of other fatty lesions in the ipsilateral or contralateral kidney, independent of the dominate lesion is a strong indicator of renal angiomyolipoma.

In this case, the large exophytic fatty tumor of the left kidney had all three distinctive CT findings as previously mentioned, allows the differential diagnosis of this condition from well-differentiated retroperitoneal liposarcoma quite clearly.

CONCLUSION

Large exophytic renal angiomyolipoma is a large benign renal tumor that extend to perirenal space. It has a characteristic fat component that can clearly be demonstrated by CT. CT accurately depicted the presence, location and size of the tumor, provided information about its relation to adjacent structures and showed the important findings that lead to accurate distinction of large exophytic renal angiomyolipoma from retroperitoneal perirenal liposarcoma.

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