[症例報告]A HUGE RENAL ANGIOMYOLIPOMA MIMICKING A RETROPERITONEAL SARCOMA : A CASE REPORT

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A HUGE RENAL ANGIOMYOLIPOMA MIMICKING A RETROPERITONEAL SARCOMA: A CASE REPORT

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Key Words: angiomyolipoma, kidney, non-tuberous sclerosis, leiomyosarcoma-like feature

Abstract

A case of renal angiomyolipoma which mimicked a retroperitoneal sarcoma and presented a diagnostic problem is reported.

The patient was a 63-year-old female who complained of right flank mass with no other accompanying symptoms. The US and CT demonstrated a huge mass in the retroperitoneum and near the lower pole of the right kidney. A selective right renal arteriogram showed stretched out feeding arteries outlining the huge tumor with tumor stains at the venous phase. An inferior venocavogram revealed a compressed segment by the tumor. At operation, the tumor was found to originate in the lower pole of the right kidney. It was oblong in shape, 16 × 15×9 cm in size and 1,800gm in weight with a massive central necrosis on cut sections. Histologically, the tumor was composed of a mixture of the mature fatty tissue, thick-walled vessel and smooth muscle. The smooth muscle cells showed pleomorphism and mitoses like leiomyosarcoma with no evidence of capsular invasion. The final pathological diagnosis was renal angiomyolipoma.

The patient has been doing well 6 months after surgery.

Introduction

Renal angiomyolipoma is a benign mesenchymal tumor which is considered to be uncommon, and thus, infrequently diagnosed preoperatively. Recent advances in the ultrasonography(US) and computed tomography(CT) have readily made the diagnosis of this tumor. However, A huge tumor with a massive necrosis may cause a problem in differentiating it from other renal and retroperitoneal tumors. Our case we recently encountered had a tremendous sized tumor with a massive necrosis and showed arteriographic and histologic features suggesting malignancy. Its rarity, the occasionally preoperative diagnostic difficulty, and the histologic features simulating malignancy prompted us to present this case with a review of the clinical and histologic features, and treatment of this rare tumor.

Case Report

A-63-year-old woman, married, multigravida was referred to the University Hospital on November 7, 1985 for further evaluation of the right flank mass. On October 1984, the patient

noticed a mass at the right flank which was approximately a chicken egg's size. Since she had no other accompanying symptoms, she did not seek any medical consultation. One year later, the mass increased up to a size of an infant's head with no other symptoms. During her clinical course, the patient denied any abdominal pain. Her obstetric, family and past history were noncontributory.

On admission, the patient was afebrile and well-nourished. Physical examination revealed the patient to be intelligent and there were no stigmata of tuberous sclerosis. Upon examination of the abdomen, a mass of 20×16 cm was felt extending from the right subcostal margin to the right superior spinous process of the ileum. It was oblong in shape, elastic firm in consistency and smooth in surface. It fixed retroperitoneally with no pulsation.

Laboratory Findings

Complete laboratory examination was performed. The pertinent findings were slight increase in LDH, positive urinary occult blood and decrease in PSP and Fishberg's concentration test.

CT and US Findings

The CT demonstrated a huge mass located in the retroperitoneum and near the lower pole of the right kidney. The CT appearance of the tumor was cystic(homogeneous, central low attenuation values) with an irregular marginal high attenuation (Fig.1). The US showed a mass with an irregular marginal high and central low echoes.

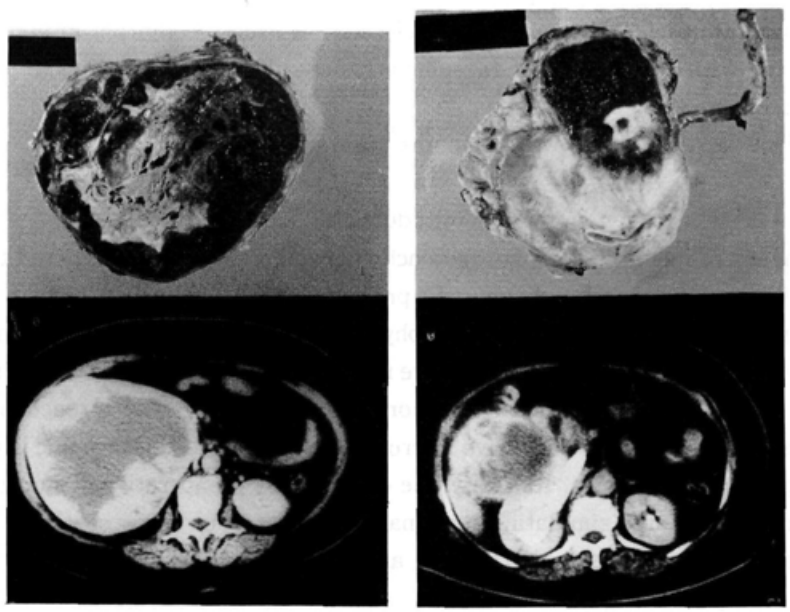

Fig.1 Photomacrographs of cross section of the tumor with its counterpart of CT scan. at the level of the greatest diameter of the tumor (left) and of the lower pole of the kidney (right).

Angiographic Findings

Selective right renal arteriogram revealed stretched out feeding arteries outlining the huge mass with tumor stains at the venous phase(Fig.2). An inferior venocavogram revealed a segment with medial compression by the mass.

Other ancillary procedures such as barium enema, barium meal upper GI series and intravenous pyelography did not reveal any significant findings.

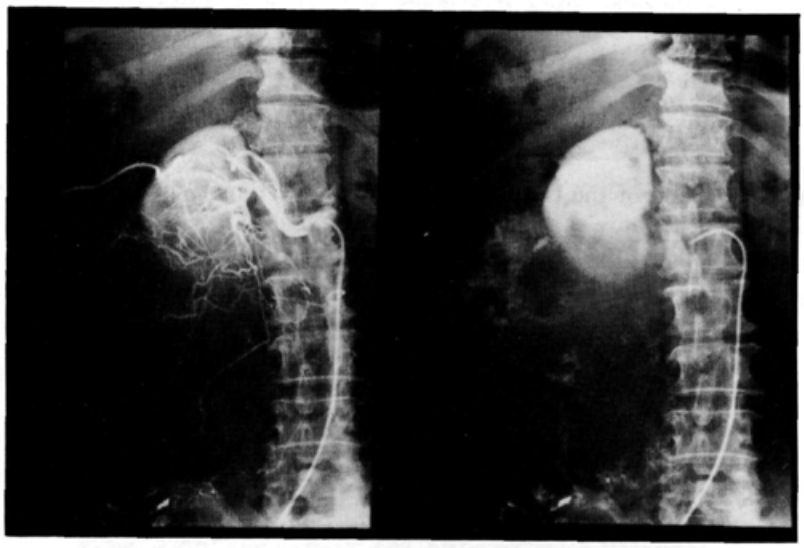

Fig.2 Selective renal arteriograms showing stretched out feeding arteries outlining the huge tumor (left) and tumor stains at the venous phase (right).

Operative Findings and Procedures

The operation was carried out on December 3, with a diagnosis of retroperitoneal sarcoma around the kidney and, to a lesser possibility, renal tumors. The tumor was located in the retroperitoneum and covered with the mesentery of the colon which was separated from the tumor with difficulty. The tumor was partially adherent to the retroperitoneum and the iliopsoas muscle posteriorly and inferiorly. Although there was a compression of the vena cava by the tumor, no adhesion or invasion was found. When the tumor was completely separated from the surrounding tissues, the tumor was found to originate in the lower pole of the right kidney. To us, the tumor appeared malignant and also, intraoperative pathological report was probably malignant. Therefore, right nephrectomy and regional nodal dissection were carried out.

Pathological Findings

The tumor was oblong in shape, 16×15×9 cm in size and 1,800gm in weight. The external surface was relatively smooth with hemorrhages. On cut sections, it appeared demarcated and

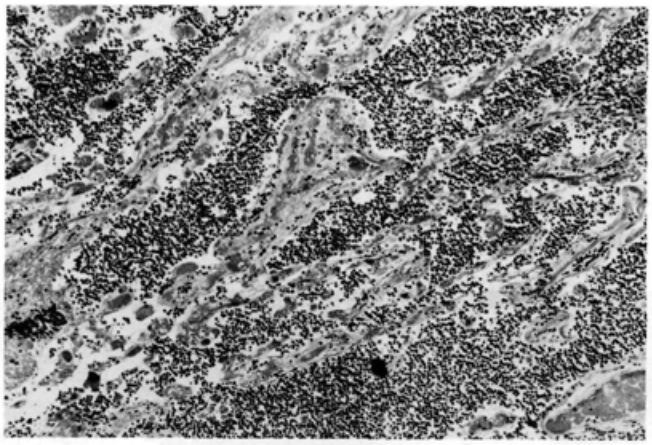

Fig.3 Photomicrograph of the hemorrhagic area of the tumor showing thick-walled vessels with red cells (HE, ×66).

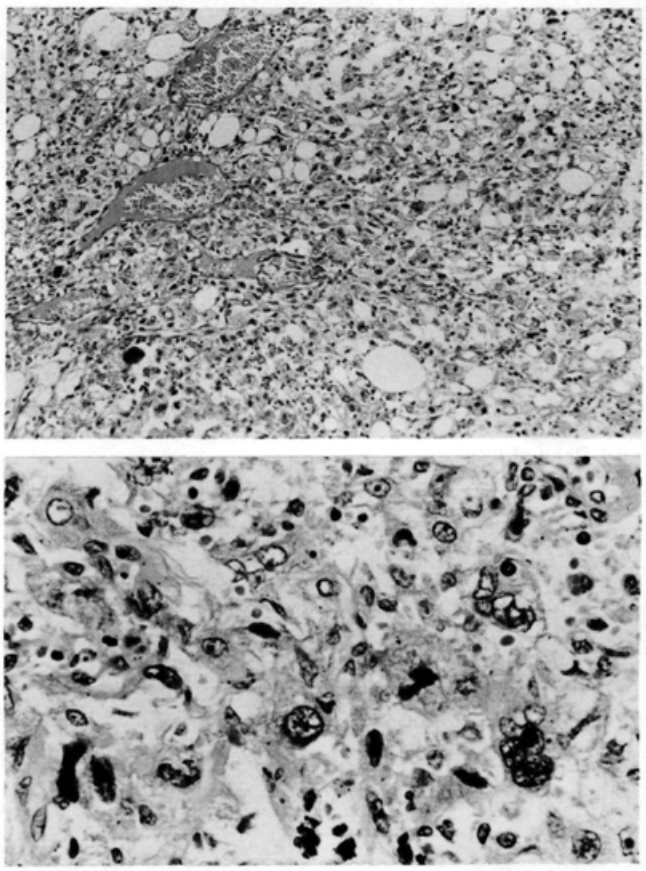

Fig.4 Photomicrographs of the tumor showing a mixture of thick -walled vessels, fat cells and pleomorphic smooth muscle cells(top; HE, ×66) and atypical, pleomorphic smooth muscle cells(bottom; HE, ×132).

capsulated with a massive hemorrhage and necrosis (Fig.1).

The multiple histologic sections revealed that the tumor was composed of a mixture of the mature fatty tissue, thick-walled vessels (Fig.3) and sheets of smooth muscle. The fatty tissue was scanty and evident near the capsule. The great majority of tumor components was the vessels and smooth muscle cells. Amongst the three tissues, the predominating tissue was the myogenic element. In areas toward the center of the tumor, the muscle cells were atypical and pleomorphic with bizarre hyperchromatic nuclei, multinucleated giant cells and mitotic activity (Fig.4). There was no evidence of capsular or vascular invasion. The final pathologic report was renal angiomyolipoma.

Discussion

Renal angiomyolipoma is a rare benign tumor composed of variable admixture of blood vessels, smooth muscle cells and mature fat cells with the predominance of one of the three elements[1]. The disease is usually associated with tuberous sclerosis but there are reported cases without tuberous sclerosis[2)-5)]. Thus, clinically the diseases are found in two forms, each with distinct features. Approximately 50%-80% of the disease is associated with tuberous sclerosis [6]. In Japan, its frequency with tuberous sclerosis is about 43%[7].

The renal angiomyolipomas associated with tuberous sclerosis are usually bilateral, small, multifocal, asymptomatic and with an equal sex distribution. On the other hand, those without it are usually unilateral, large, unifocal with a sex distribution towards the female, and occurring most commonly during the fourth or fifth decades of life [8]. A peculiar feature of the solitary angiomyolipoma is that it has a predilection to occur in the right side, accounting for 80% of the solitary tumor[9]. The renal angiomyolipoma in this case was not associated with tuberous sclerosis and was a large solitary tumor in the right side.

The diagnosis may be readily made when it is associated with tuberous sclerosis. However, when a solitary renal tumor is found without the stigmata of tuberous sclerosis, the disease is quite difficult, even impossible, to diagnose. Neverthless, there are several reports that has described the possibility of the preoperative diagnosis with such features as marked increase of LDH, marked radiolucent area in the film of the abdomen, hypervascularity in angiography, low attenuation values in CT, and inner mosaic pattern in US [10),11)]. Yet, the features described above may also be seen in cases of renal cell carcinoma, lipoma and liposarcoma, so that some authors still hold that the diagnosis is not accurate preoperatively[12)-18)]. At least in this case, because of the low content of the fatty tissue, massive necrosis and hemorrhage and the predominance of the myogenic element, the diagnostic features described above were not found leading us to suspect more of the retroperitoneal sarcoma.

With regards to the size of the tumor, the review of the literature in Japan has revealed that the largest tumor reported so far is 1,800gm. in weight [19)], followed by 1,700gm.[20)]. The tumor in this case weighed 1,800gm. Such huge tumor is rarely encountered and when found as a large solitary tumor, it usually poses difficulty in diagnosis before surgery and even at surgery.

Concerning the treatment of renal angiomyolipoma, until recently most patients underwent radical nephrectomy since most of the tumors were not correctly diagnosed until after the renal

tumor was removed. Nowadays, it is generally accepted that since renal angiomyolipomas are clinically benign, nephrectomy is not required by their presence alone. Thus, surgery should now play a minor role in treatment of renal angiomyolipoma. Partial nephrectomy or enucleation[21)-23)], therapeutic embolization[24)-26)], and mere follow-up[27),28)] for preservation of the remaining kidney tissue are recommended. However, in practice, the biggest problem regarding treatment remains in those cases of a huge solitary tumor not associated with tuberous sclerosis, of a tumor with local invasion or massive hemorrhage, and in whom the presumptive diagnosis will be malignant[29)-31)]. In these cases like our case with a huge tumor, surgery may play a major role in treatment. In our case, since the preoperative diagnosis was not definitive and it was impossible even at surgery and on histologic study during operation, we opted to perform nephrectomy.

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