Renal angiomyolipoma with renal vein invasion

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Renal angiomyolipoma is a uncommon benign tumor, considered an hamartoma. The lesion, usually benign, can be single or multiple and well-circumscribed. In letterature only few cases of infiltrating angiomyolipomas have been described. The aim of the paper is to describe a paradigmatic case of a giant kidney angiomyolipoma, not associated with tuberous sclerosis, invading the pelvis and the renal vein. The lesion have been discovered incidentally during abdominal ultrasound for other pathology. Owing to the extent of the lesion and the appreciable risk of bleeding, we opted for surgical treatment.

KEY WORDS: Angiomyolipoma; Kidney; Renal vein invasion; Radical nephrectomy.

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INTRODUCTION

Angiomyolipoma is a nodule composed of variable amounts of mature adipose tissue, smooth muscle, and thick-walled blood vessels derived from perivascular epithelioid cells usually arising in the renal cortex. Its prevalence in the general population has been reported to be 0,3-3% overall in the female patients (1). In the multifocal form, it is usually associated to tuberous sclerosis (2). The average lesion size is from 2 mm to 20 cm maximum diameter. In most cases the angiomyolipoma is asymptomatic and is diagnosed incidentally with Ultrasound, CT and MRI done for other reasons. Renal angiomyolipoma at times can be aggressive and may show extension into renal vein and inferior vena cava (3). We describe a paradigmatic case of a giant kidney angiomyolipoma, not associated with tuberous sclerosis, invading the pelvis and the renal vein.

CASE REPORT

The lesion have been incidentally discovered in a 78 years old woman by ultrasound scan done for other reasons. Total body CT scan and MRI have been done showing large node (18 mm) in the mediastinus, a 8 cm large lesion of the upper pole of the left kidney with prevalence of fat tissue and solid areas invading the renal vein for 4 cm, some large nodes (1 cm) in the retroperitoneum and gallbladder stones (Figure 1, 2). Past medical history included breast reduction; hyatal hernia surgery; hypothiroidism; pulmunary infection during the last months. Blood tests: Hb 15 g/dl-1 leukocyte 8 x 103 µl-1, urea 44 mg/d-1, creatinine 0.9 mg/dl-1. The extension of the lesion, the risk of bleeding and the risk of renal carcinoma were carefully evaluated in order to decide the surgical treatment. The patient underwent laparoscopic left adrenal sparing radical nephrectomy. Definitive histology: renal angiomyolipoma invading the renal vein with negative ilar nodes and normal left renal parenchima (Figure 3, 4). No post-operative complications. The patient was discharged in the fifth postoperative day. At the first control, one month after the operation, the patient was asymptomatic, the abdominal ultrasound scan was normal and the blood tests were normal.



Figure 1. Axial CT section showing extension of the lesion into the renal vein.

Figure 2.

T2 weighted MRI mass shows hyperintense signal with extension into renal vein.

Figure 3.

Renal angiomyolipoma: blood vessels with thickened walls mature adipose tissue and smooth muscle cells mixed. EEx20





Figure 4. Renal

angiomyolipoma: renal vein invasion. EEx5

DISCUSSION

Renal angiomyolipoma is usually a benign lesion, it can be rarely associated to renal adenocarcinoma (4, 5) and to tuberous sclerosis. The histology shows mature adipose tissue, smooth muscle, and thick-walled blood vessels. The sporadic angiomyolipoma is monolateral and more frequent in the females. When associated to tuberous sclerosis, angiomyolipomas are multiple, bilateral and larger. The clinical interest in angiomyolipoma is in its rapid growth, the difficulty in distinguishing it from malignant lesions, the difficulty of establishing the diagnosis and correct treatment.

The diagnosis is usually made by ultrasound scan that find a hyperecogenic omogeneus lesion in the renal cortex. The large lesions may have disomogeneus areas that make the diagnosis difficult; TC scan is usually mandatory in the large lesions: the diagnosis is based on the identification of fat inside the lesion (6). MRI is useful only in some complex cases (7). Lesion larger than 4 cm may bleed, may cause flank pain and may be palpable (8).

When the lesion is growing, when it is symptomatic or when the differential diagnosis is doubtful, surgical treatment is necessary: enucleoresection, embolization or radical nephrectomy (9).

CONCLUSIONS

In most cases angiomyolipoma is asymptomatic and it is an incidental finding during Ultrasound scan or CT scan done for other reasons. It may involve regional nodes, renal vein or inferior vena cava, that can suggest an aggressive evolution (10); anyway, these lesions are not considered metastasis. In case of benign lesion the treatment has to be conservative. Radical surgery is requested in those rare cases where the angiomyolipoma is really large or involves the renal vein.

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