ANGIOMYOLIPOMA OF THE KIDNEY: REPORT OF 12 UNILATERAL CASES WITHOUT TUBEROUS SCLEROSIS AND REVIEW OF THE LITERATURE

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ÖZET

Angiomyolipomlar yağ dokusu, atipik kalın duvarlı damarlar ve düz kastan oluşan selim renal neoplazmalar olarak kabul edilir. Çoğu tüberoz skleroz ile birlikte görülür. Son 20 yıl içinde fakültemizde 12 tane unilateral ve tüberoz skleroz bulunmayan renal angiomyolipom vakası teşhis edilmiştir. Kadın erkek oranı 5:1'dir. Sekiz hastaya nefrektomi, dört hastaya da sadece tümör enükleasyonunu içeren böbreği koruyucu elektif bir operasyon uygulanmıştır. Üç vakada tümörün multinodüler oludğu görülmüştür. Bir hastada da sürrenalde tümöral bir kitle tesbit edilmiştir. Mikroskopik olarak; bir hastada hiperplastik damarsal yapılar, iki vakada düz kas ve dokuz vakada yağ dokusunun baskın histolojik yapı olduğu görülmüştür. Hiç bir hastada diğer böbrekte tümoral bir yapıya rastlanmamıştır. Hernekadar angiomyolipomlardan renal cell karsinom geliştiği ya da olayın tamamen bir tesadüf olup olmadığı kesin değilsede renal angiomyolipomlar ile renal cell karsinom arasında bir bağlantı olduğunu belirten bazı yayınlar mevcuttur. Bu nedenle biz de bu hastalarda renal parankimi koruyacak lokal bir rezeksiyonu içeren cerrahi eksplorasyonu tavsiye etmekteyiz.

SUMMARY

Angiomyolipoma has been accepted as a benign renal neoplasm composed of mature adipose tissue, atypical thick-walled blood vessels and smooth muscle. Most of theme are associated with tuberous sclerosis. In the last 20 years the diagnosis of unilateral renal angiomyolipoma without tuberous sclerosis was made in 12 patients in our institution. Female-to-male ratio was 5;1. Eight of the patients underwent nephrectomy and in four cases elective exploration and a renal preserving operation by only enuclation of the tumor were applied. The tumors were multinodular in three cases. In one case, a tumoral mass was also observed in the adrenal gland. Microscopically; in one case hyperplastic vessels, in two cases smooth muscle and in nine cases mature adipose tissue were the prominent components. Involvement of the controlateral kidney has not been noted in any of our patients. Although it has to be determined whether it is coincidental or renal cell carcinoma originates in an angiomyolipoma there are several reports indicating an association between renal angiomyolipoma and renal cell carcinoma. So we recommend surgical exloration but a concervative local resection with preservation of the renal parenchyma.

INTRODUCTION

The term renal angiomyolipoma was first used by Morgan and associates in 1951 (1). It is an uncommon benign tumor and an unusual pathological entitiy with a large clinical spectrum. It may be an asymptomatic isolated unilateral hamartoma or massive bilateral renal involvements in conjunction with tuberous sclerosis. More than 200 cases of angiomyolipomas without tuberous sclerosis have been reported (2,3). Renal angiomyolipomas with or without tuberous sclerosis are hamartomatous and benign in nature. 50 % of all renal angiomyolipomas occur in patients with tuberous sclerosis (4,5,6) and 40 - 80 % of tuberous sclerosis patients exhibit angiomyolipomas (7). In the diagnosis of renal angiomyolipomas, plain radiographs of the abdomen showing a renal mass of variable density, angiography revealing a hypervascular mass, ultrasound demonstrating the hyperechocenity due to lipid density and CT

demonstrating decreased attenuation units (negative Hounsfield units) confirming the presence of fat in the tumor, are the usefull methods of choise.

PATİENT AND METHODS

Twelve cases of unilateral renal angiomyolipomas without tuberous sclerosis are reported. Of the 12 patients 10 were female and 2 were male with a ratio of 5;1. Patients age ranged from 22 to 60, mean age 52.1 years. Of these patients 5 presented with urinary infection, micro hematuria and flank pain (41.7 %), 3 with palpable mass (25 %), 3 with microhematuria (25 %) and 1 case diagnosed incidentally (8.3 %) during a rutin examination. No patient presented with the classical triad of mass, flank pain and hematuria. Excretory urography showed a renal mass in 9 of the 12 patients. Ultrasonography was positive for all patients showing the hyperechoic characteristics of angiomyolipoma (Fig 1,2). CT was performed in the last 8 patients which revealed tumors with lipid density due to decreased attenuation units (Fig 3,4,5). A correct preoperative diagnosis was made in 8 patients (66.6 %) with IVP, ultrasound and CT, in 3 (25 %) with ultrasound and in 1 patient (8.3 %) with I.V.P. and angiography. Eight patients underwent nephrectomy and 4 were treated by the local excision of the tumor. There were no operative death and no important postoperative complication.

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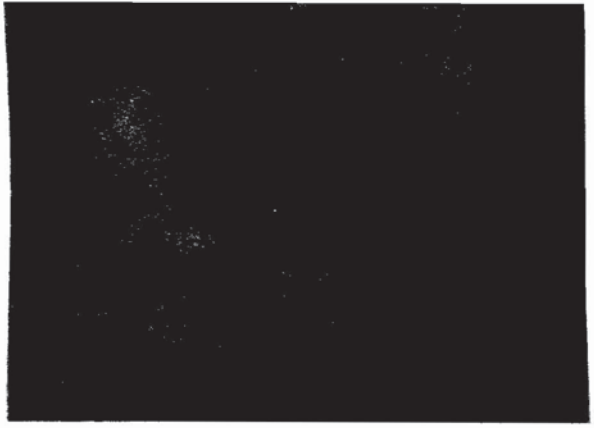

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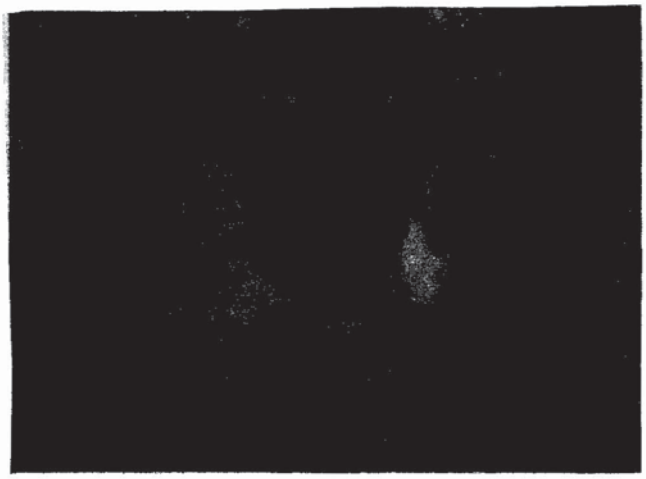

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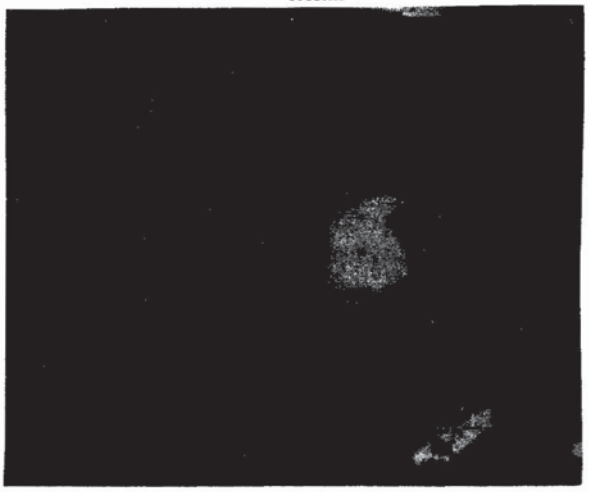

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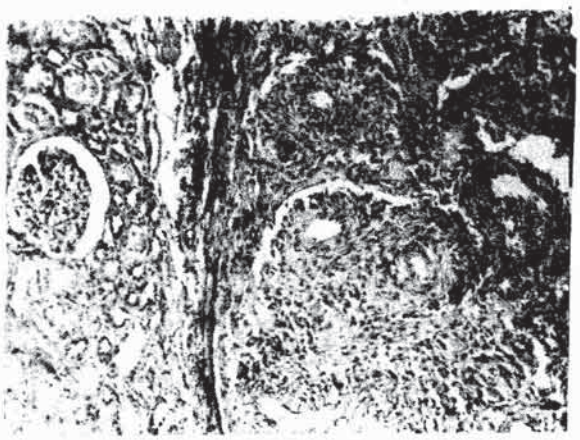
RESULTS

In 5 cases the tumor was in the right and in seven in the left side. The size of the tumors ranged from 3 to 40 cm. with a mean of 10.5 cm. Mostly they were round, some nodular and predominantly yellow in apperance. In 4 cases tumors were limited within the renal capsule and located in the cortex of the kidney. Multiple tumor masses were found in 3 cases. I 5 the tumor invaded the renal capsule and renal pelvis. In ona case lipoid mases were present in the adrenal gland and liver. Microscopically in all cases, the vascular comporent of the tumors consisted of hyperplastic, thick walled tortuous blood vessels. The vessels were surrounded by cllars of smooth muscle cells which extended as strands into adipose tissue. With the application of Van Gieson elastic dye it was observed that the vessels did not have lamina elastica interna. In one case the only prominent component was the hyperplastic vessels.



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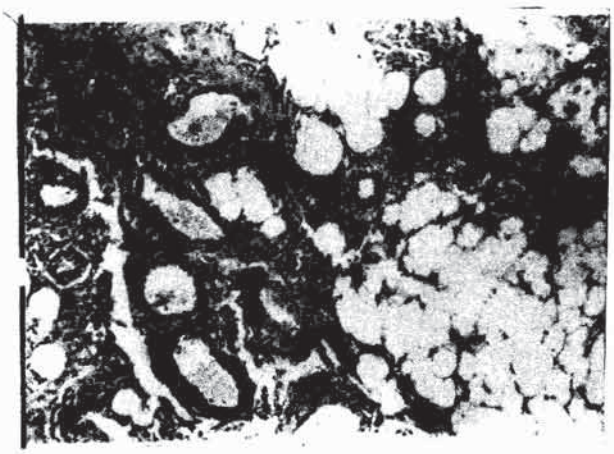

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The adipose tissue was composed of uniform fat cells with a large cytoplasmic vacuoles and a small peripherial nucleus (Fig 6,7). There were slight cellular pleomorphism in 3 cases and moderate in 2. But neither conciderable pleomrphism nor mytosis were observed. In renal tissue next to tumor, glomerular sclerosis, tubular atrophy and interstitiel fibrosis were also noted.

DISCUSSION

The term angiomyolipoma which was first used by Morgan has been generally referred to as a hamartoma. The association of renal angiomyolipomas with tuberous sclerosis has been known for more than 100 years, since Bourneville first noted the lesions in autopsy speciments (8). However more than 200 cases have been reported in which there has been no evidence of coexistent tuberous sclerosis. Renal angiomyolipomas are 4 times as common in female than is male and tent do be unilateral (3). In our cases the female-tomeal ratio was also 5:1, and all of them even the small ones were unilateral contrary to the belief that small renal nagiomyolipomas are bilateral. The size of renal angiomyolipomas may vary from a few milimeters to many centimeters. In our cases the size of the tumors ranged from 3 - 40 cm. The belief that angiomyolipomas without tuberous sclerosis are large and sympotamitc is changing. In a recent review of the literature Oesterling and ass. found that the majority (77 %) of lesions were 4 cm. or less and asympotamitc. In accordance with his observation the tumors were smaller than 4 cm. in 4 of our cases. Clinical presentations of angiomyolipomas in patients with and without tuberous sclerosis is variable. Five of our patients presented with urinary infection microhematuria and flank pain, three with only a palpable mass, three with microhematuria, and one case was diagnosed incidentally.

Price and Mostofi emphasized the importance of hemorrhage and 30 percent of the cases that they reviewed presented as acute surgical emergencies (9). In our study like in Price and Mostofi's research, intralesional hemorrhage was found in 5 cases and an extrarenal hematoma in one case.

The very important point in renal angiomyolipomas is to differentiate them from renal cell carcinoma. By means of CT scan, renal ultrasound and angiographic criteria renal angiomyolipomas can be identified. The hyeperechogenicity in ultrasonography and low attenuation coefficient in CT scan indicates angiomyolipoma. Some reports emplasize the association between renal angiomyolipomas and renal cell carcinomas (10,11,12,13). It is not clear whether the carcinoma orginates in an angiomyolipoma or it is a coincidental finding.

Hajde and Foote reported 14 cancers in 53 patients with renal tumors and tuberous sclerosis (14), but there is only 4 cases of angiomyolipoma and coincidental renal cell carcinoma in patients without tuberous sclerosis reported in the literature (15,16). Blate and ass. added 2 more cases o coincidental cancer with surgically proved isolated angiomyolipoma (17).

So the prognosis of angiomyolipoma is variable and it is very difficult to predict by size or other means present at the begining. We believe that for the benefit of the patient every angiomyolipoma, small (3 cm.) or large (3 cm.), simple or complex (without classical CT and ultrasonographic characteristics) must be surgically treated. But elective exploration and preservative surgery, if possible must be the method of choice.

In recent reports many authors like Malone and Blute (2,17) conservative surgical resection (especially in tumors larger than 3 cm.) with preservation of renal parenchyma is advised in accordance with our application.

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