GIANT RENAL ANGIOMYOLIPOMA – CASE REPORT

*Konrad Wroński^{1, 2}, Zbigniew Masłowski²

¹Department of Oncology, Faculty of Medicine, University of Warmia and Mazury, Olsztyn, Poland Head of Department: prof. Sergiusz Nawrocki, MD, PhD ²Department of Surgical Oncology, Hospital Ministry of Internal Affairs with Warmia and Mazury Oncology Centre, Olsztyn, Poland Head of Department: Andrzej Lachowski, MD

Summary

Renal angiomyolipoma is rare benign renal neoplasm composed of variable amounts of adipose tissue, smooth muscle tissue originating from perivascular epithelial cells and blood vessels. The incidence of renal angiomyolipoma is from 0,3% to 3,0%. Tumors over 10 cm are diagnosed rare. Clinically renal angiomyolipoma is usually associated with tuberous sclerosis complex and is more frequent in women. About 20% of these tumor are seen in patients with the tuberous sclerosis complex and 80% patients with tuberous sclerosis complex develop renal angiomyolipomas. The tumor is diagnosed by ultrasound, computer tomography scan (CT), magnetic resonance imaging (MRI), histology and immunohistochemistry.

The authors of this article presented a case of a 47-year-old Caucasian woman who was admitted to the hospital because of giant renal angiomyolipoma. The patient underwent radical left nephrectomy without complications. The authors performed a literature review on giant renal angiomyolipoma treatment.

Key words: angiomyolipoma, kidney neoplasm, giant kidney tumor, treatment, surgery

INTRODUCTION

Renal angiomyolipoma is rare benign renal neoplasm composed of variable amounts of adipose tissue, smooth muscle tissue originating from perivascular epithelial cells and blood vessels (1-3). The incidence of renal angiomyolipoma in general population is less than 0.2% and prevalence from 0.3% to 3.0% of all surgical resected renal tumors (3). The average size of these tumors is 2 to 8 cm and there are rarely diagnosed tumors over 10 cm (4).

CASE REPORT

A 47-year-old Caucasian woman was referred to the Department of Surgical Oncology due to giant left kidney tumor diagnosed in computer tomography scan (fig. 1, 2). Computer tomography scan was typical for renal angiomyolipoma size 141 x 100 x 168 mm. CT scans showed also several small tumors in the lower pole of the left kidney and left adrenal adenoma. Left adrenal tumor was not hormonally active.

The patient suffered from abdominal pain located in the left upper quadrant and constipation. Physical examination showed enlarged left kidney with palpable tumor. There were no peritoneal symptoms. Blood tests were normal. There was no histopathological examination of the fineneedle aspiration biopsy before planned surgery.

She had no any other symptoms, there was no history of weight loss and loss of appetite. She had two surgeries – first appendectomy due appendicitis and excision of uterine polyp. There was no history of carcinoma in patient family. The patient was taken to the operating room for an open left nefrectomy. During the surgical exploration claimed giant left kidney angiomyolipoma with rich network of vessels. Left kidney with giant tumor and adrenal adenoma were removed (fig. 3, 4). The material was sent for routine histopathological examination. The time of surgery was about 90 minutes.



Fig. 1. Computed tomography scan suggesting giant renal angiomyolipoma.



Fig. 2. Computed tomography scan showing giant pathological mass in left kidney measuring 141 x 100 x 168 mm.



Fig. 3. Specimen showing giant pathological mass in the left kidney.

Histopathology and immunohistochemistry examination showed mature adipocytes, blood vessels and smooth muscles without neoplasm component. Patient after surgery felt good and did not complain of pain. The postoperative period was uncomplicated and the patient left the ward in the 6th day after surgery.

DISCUSSION

Angiomyolipoma is a rare benign soft tissue tumor involving kidney, liver or other visceral organs. Clinically renal angiomyolipoma is usually associated with tuberous sclerosis complex and is more frequent in women. About 20% of these tumor are seen in patients with the tuberous sclerosis complex and 80% patients with tuberous sclerosis complex develop renal angiomyolipomas (5).

The tumor is diagnosed by ultrasound, computer tomography scan (CT), magnetic resonance imaging (MRI), histology and immunohistochemistry. The presence of



Fig. 4. Giant renal angiomyolipoma.

fat on computer tomography imaging inside renal lesion give us information about renal angiomyolipoma (6). When on CT scan there are more than 20 pixels with attenuation less than -20HU and of more than 5 pixels with attenuation less than -30 HU renal angiomyolipoma can be indentified (6). Histopathological examination shows variable amounts of adipose tissue, smooth muscle tissue originating from perivascular epithelial cells and blood vessels (1-3). Immunohistochemical examination of tumor by beta-hydroxy beta-methylbutyric acid-45 (HMB-45) and CD-68 positivity combined with cytokeratin negativity are important parameters for correct diagnosis (7, 8). Renal angiomyolipoma must be correct differentiated from renal cell carcinoma and liposarcoma.

The management of renal angiomyolipoma is based on tumor size, risk stratification for bleeding and malignancy suspected. Tumors which size is greater than 4 cm should be operated (4). The risk of hemorrhage is related to the tumor size and is significant greater than tumor is larger than 4 cm (9).

For tumor larger than 4 cm surgical intervention should be considered. Patients in whom preservation of renal function is important and tumor is not so big size, a nephron sparing surgery, partial nephrectomy or selective transcatheter arterial embolization can be performed. In case of angiomyolipomas larger than 8 cm open surgery is preferred. Selective transcatherter arterial embolization is safe procedure which can be done to decrease the risk of bleeding without loss of renal function and is recommended to avoid excess blood loss during surgery (10, 11). Nephron sparing surgery is an acceptable type of treatment, alternative to selective arterial embolisation (12, 13). It is possible to treat patients with renal angiomyolipoma by laparoscopic cryoablation as an minimal invasive procedure (14).

7

References

1. Faraji H, Nguyen BN, Mai KT: Renal epithelioid angiomyolipoma: a study of six cases and meta-analytic study. Development of criteria for screening the entity with prognostic significance. Histopathology 2009; 55: 525-534. **2.** Schieven LW, Smedts F, Hopman AH et al.: Fine needle aspiration using improved agar microbiopsy is highly concordant with renal mass final diagnosis and subclasification. J Urol 2009; 182: 2590-2593. **3.** Zhang JQ, Fielding JR, Zou KH: Etiology of spontaneous perirenal hemorrhage: a meta-analysis. J Urol 2002; 167: 1593-1596. **4.** Nelson CP, Sanda MG: Contemporary diagnosis and management of renal angiomyolipoma. J Urol 2002; 168: 1315-1325. **5.** Earthman WJ, Mazer MJ, Winfield AC: Angiomyolipomas in tuberous sclerosis: subselective embolotheraphy with alcohol, with long-term follow-up study. Radiology 1986; 160(2): 437-441. **6.** Vitaly M, Surena FM, Christopher GW: Benign renal tumors. [In:] Campbell-Walsh Urology. 10th ed., Saunders Elsevier, Philadelphia 2012: 1498-1501. **7.** Svec A, Velenska Z: Renal epitheloid

Received: 26.01.2015 Accepted: 18.02.2015

angiomvolipoma - a close mimic of renal cell carcinoma. Report of a case and review of the literature. Pathol Res Pract 2005; 200: 851-857. 8. Kulkarni B, Desai SB, Dave B et al.: Renal angiomyolipomas - a study of 18 cases. Indian J Pathol Microbiol 2005; 48: 459-463. 9. Gaikwad AB, Madathil MB, Kothari AS: Giant renal angiomyolipoma with fatal hemorrhage due to a large pseudoaneurysm. J Clin Ultrasound 2008; 36: 174-176. 10. Singla A, Chaitanya Arudra SK, Bharti N: Giant sporadic renal angiomvolipoma treated with nephron-sparing surgery. Urology 2009; 74: 294-295. 11. Bishay VL, Crino PB, Wein AJ et al.: Embolization of giant renal angiomyolipomas: technique and results. J Vasc and Intervention Radiology 2010; 21(1): 67-72. 12. Mullerad M, Hidas G, Kastin A et al.: Nephron sparing surgery as a treatment modality for renal angiomyolipoma. Harefuah 2005: 144: 619-621. 13. Heidenreich A. Hegele A. Varga Z et al.: Nephron sparing surgery for renal angiomyolipoma. Eur Urol 2002; 41: 267-273. 14. Byrd GF, Lawatsch EJ, Mesrobian HG et al.: Laparoscopic cryoablation of renal angiomyolipoma. J Urol 2006; 176: 1512-1516.

> Correspondence to: *Konrad Wroński General Surgery Consultant Department of Surgical Oncology Faculty of Medicine University of Warmia and Mazury al. Wojska Polskiego 37, 10-228 Olsztyn, Poland tel./fax:+48 505-818-126 e-mail: konradwronski@wp.pl