doi: 10.5146/tjpath.2012.01102

Myxoma of the Renal Sinus: Case Report and Literature Review

Renal Sinüsün Miksoması: Olgu Sunumu ve Literatürün Gözden Geçirilmesi

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ABSTRACT

Myxoma is a rare mesenchymal tumor and it is mainly seen in heart and skin. Renal myxoma is extremely rare. To date, eleven cases of kidney myxomas have been reported in the literature. One of them is myxoma of the renal sinus. Our case was an 82-year-old man admitted to our hospital symptoms related to the urinary tract obstruction. Abdominal computerized tomography revealed a solid, hypodense mass 9 cm in diameter infiltrating the renal parenchyma in the renal pelvis. The patient underwent nephrectomy. The resected kidney contained gelatinous tumor with indistinct borders. The tumor was composed of slender, bland, spindle-shaped cells with large amounts of mucoid material. Tumor cells were positively stained with vimentin, focally stained positive for smooth muscle actin and had negative reactivity for S-100 protein, epithelial membrane antigen and pancytokeratin. Herein we report the second case of renal myxoma arising from the renal sinus.

Key Words: Myxoma, Kidney, Renal neoplasms

ÖZ

Miksoma nadir görülen benign mezankimal tümör olup literatürde sıklıkla kalp ve deride rapor edilmiştir. Böbrekte görülen mezankimal tümörler içerisinde miksoma oldukça nadir izlenmekte olup literatürde bugüne kadar 11 olgu bildirilmiştir. Bunlardan bir tanesi renal sinüs yerleşimlidir. 82 yaşında erkek hasta, parsiyel üriner obstrüksiyon semptomlarıyla hastanemize kabul edildi. Yapılan abdominal bilgisayarlı tomografide sol böbrek hilusunda yerleşim gösteren hipodens, parankimle sınırları net olarak seçilemeyen, 9 cm boyutunda solid kitle tespit edildi. Böbrekte kitle nedeniyle sol renal nefrektomi yapılan hastanın gross incelemesinde, renal sinüs kaynaklı sınırları net olarak seçilemeyen jelatinöz görünümde tümör dokusu izlendi. Mikroskopik olarak bol mukoid materyal içerisinde dağılmış, iğsi sitoplazmalı, bazıları yıldızsı görünümde hücrelerden oluştuğu gözlendi. Tümör hücreleri vimentin ile pozitif, düz kas aktini ile fokal pozitiflik gösterirken, S-100 protein, epitelyal membran antijeni ve pansitokeratin ile negatif boyanma göstermiştir. Biz burada renal sinüs kaynaklı ikinci olguyu sunduk.

Anahtar Sözcükler: Miksoma, Böbrek, Renal tümörler

INTRODUCTION

Renal pelvic tumors represent 5-10% of all the renal tumors. Almost 90% of these are transitional cell carcinomas (1). Mesenchymal neoplasms arising in renal pelvis are fairly uncommon. Most of these neoplasms originate from vascular or smooth muscle tissue and eighty-one cases of benign neoplasms and 40 sarcomas were identified in the literature.

A classification of reported mesencymal neoplasm is presented in Table I. Although benign mesencymal tumors are more frequently located in renal pelvis, malignant tumors are located in ureter (2).

Myxomas are relatively uncommon soft tissue neoplasms. This tumor can be found in various sites in the body including the skin, heart, soft tissue, head and neck. Myxoma of the kidney is extremely rare. To date, eleven cases of pure myxoma reported in the literature (Table II). Only one of them originated in renal pelvis and others were situated in renal parenchyma and capsule. Our case is the second renal myxoma arising in the renal sinus.

CASE REPORT

An 82-year-old male patient was admitted with the complaints of dysuria, flank pain and urinary obstruction for two years. He had been treated with the diagnosis of tuberculosis and had no urinary system complaints before. His physical examination was unremarkable. Laboratory values were within the normal limits. Abdominal USG revealed a mass, 9 cm in diameter, solid with heterogeneous

Received : 26.08.2010
Accepted : 19.11.2010

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echo in the left renal pelvis. CT scan showed a low density mass (Figure 1). Fine needle aspiration was performed and resulting suspicious cytology for malignancy. Patient underwent a left radical nephrectomy.

Nephrectomy specimen weighed 330 g together with perirenal fat tissue. On sagittal section, the pelvis of the left kidney contained a 9x6x5 cm semitranslucent, gelatinous tumor mass. Tumor involved the renal parenchyma. Other pathologic findings such as calculi, hidronephrosis were not detected on gross examination of the renal tissue (Figure 2). Microscopically, the tumor was composed of hypocellular and hypovascular myxoid stroma. Tumor cells were spindle, fibroblast like, oval and stellate shaped. Nuclear atypia and mitosis were not demonstrated (Figure 3). Tumor was invading renal parenchyma. The tumor and renal parenchyma were indistinct. Ureter was intact. There was no pathological finding in non-tumoral areas. The myxoid stroma was stained with alcian blue and tumor cells were positively stained with vimentin (Figure 4), focally stained positive with smooth muscle actin (SMA), and negatively stained for S100 protein, epithelial membrane antigen (EMA), and pancytokeratin. Final diagnosis was myxoma of the renal sinus according to gross, microscopic and immunohistochemical findings.

Table I: Classification of mesenchymal neoplasms of renal pelvıs and ureter

Benign	Malignant
Leiomyoma Hemangioma Lymphangioma Neurofibroma Solitary Fibrous Tumor Schwannoma Glomus tumor	Leiomyosarcoma Malignant Schwannoma Angiosarcoma Osteosarcoma Chondrosarcoma Rhabdomyosarcoma

Table II: Clinicopathologic features of reported cases

Authors	Age/Sex	Site/Location	Symptom	Tumor size (cm)	Year imagine studies	Treatments	Reference No:
Appel	No /no	Right parapelvic	Hematuria for 2 months	8 cm	No	Enucleation of mass	14
Shenansky	62/male	Right Lower pole	Hematuria for 6 months	4 cm	No	Nephroctomy	15
Melamed	52/female	Left Lower pole	Renal colic	7 cm	No	"	7
Melamed	68/female	Right Upper pole	Asymptomatic	10 cm	No	"	7
Kundu	36/male	Left/mass of renal parnkima	Hypocon-drium mass for 2 months	28 cm	No	"	8
Val Bernal	37/male	Right capsule	Asymptomatic	6 cm	No	"	9
Owari	62/ male	Right Middle portion of the kidney	Asymptomatic	8 cm	No	"	10
Nishimoto	No/no	No	No	No	1996	No	11
Nishimoto	36/male	Left Lower pole	Asymptomatic	9 cm	2007	Nephroctomy	13
Bolat F	27/female	Left Lower pole	Asymptomatic	15 cm	2007	"	3
Present case	82 male	Left Renal sinus	Dysuria, urinary obstruction and flank pain	9 cm	2007	"	
Koike	No	No	No	No	No	No	12

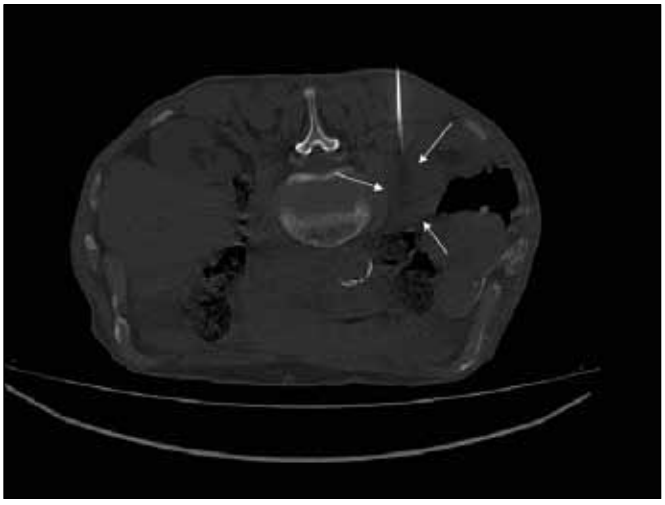

Figure 1: Computed Tomograhy image without contrast media in prone position showing hyperdense biopsy needle entering hypodense mass in the left kidney.

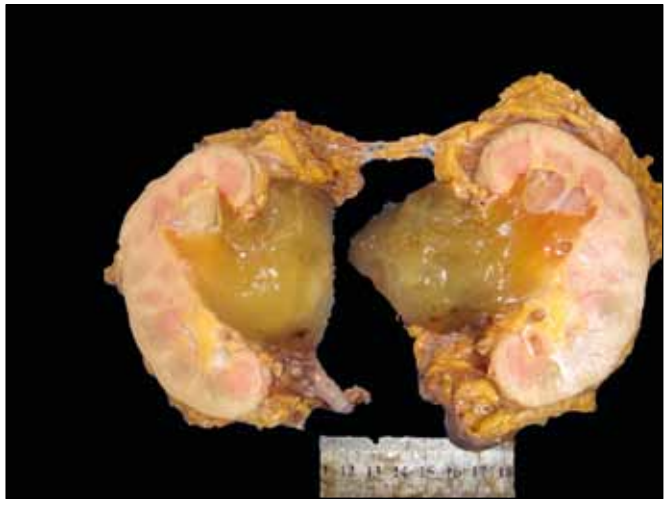

Figure 2: Tumor tissue 9 cm in diameter demonstrated in the left kidney. The surface of the greenish mass had a semitranslucent appearance and the mass was composed of a gelatinous, stringy mucoid material.

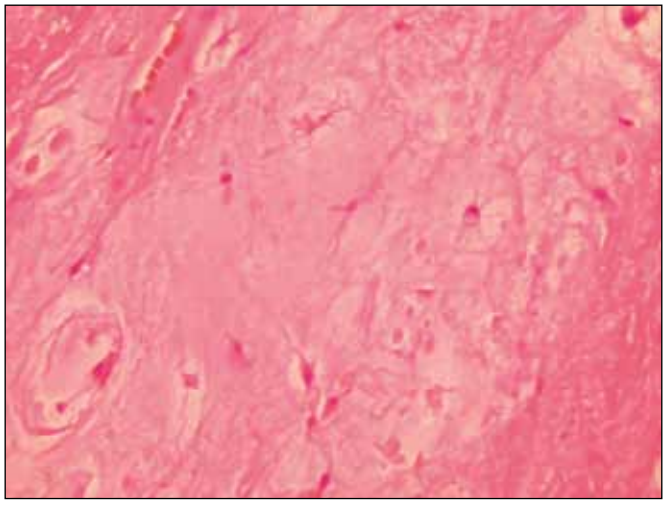

Figure 3: Photomicrograph shows tumor tissue including scattered stellate cells in the myxoid material (H&E, x200).

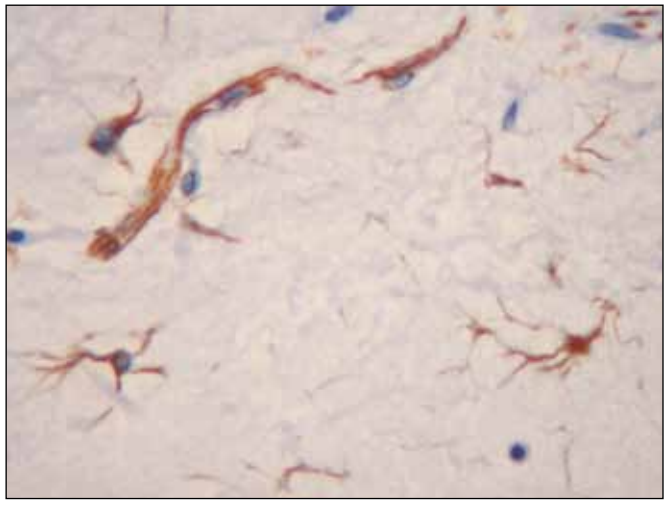

Figure 4: Tumor cells positively stained with vimentin (x400).

DISCUSSION

Various benign mesenchymal tumors of the renal pelvis have been reported in the literature (1-3). Some of these are hemangioma (40 cases), leiomyoma (10 cases), schwannoma-neurofibroma (2 cases), solitary fibrous tumor (2 cases), glomus tumor (1 case). Malignant mesenchymal tumors composed approximately 30% of mesenchymal neoplasms arising in renal pelvis (1-3). A total of 38 malignant mesencymal tumor cases have been reported in the literature up to date. Ninety percent of these are smooth muscle origin. These include leiomyosarcoma (12 cases), malignant schwannoma (1 case), angiosarcoma, osteosarcoma, chondrosarcoma and rhabdomyosarcoma (1-3). Myxoma has not been situated in classification of mesenchymal tumors of the kidney in textbooks yet.

Myxoma has been described first by Virchow in 1863 (4-6). Although myxomas show similar microscopic appearance, they exhibit different behavioral and clinicopathological course (4-6). It is suggested that this difference is based on molecular abnormalities (4-6).

Myxomas located in different sites of the body have different molecular mutations and other abnormalities (4-6).

Cells of myxoma considered to be originated from fibroblast like primitive mesencymal cells. These cells are similar to fibroblast but they lost the capacity to polymerize

collagen. Some authors believe that myxoma is a myxoid change of some mesenchymal tumors such as leiomyoma and degenerative changes seen in adipose tissue in brown atrophy of the heart (7-9). The others believe that the uniform cellular component throughout the lesion supports a neoplastic nature rather than a regressive change within the pre-existing tumor (7-9).

Clinicopathological data of the reported twelve cases are shown in Table 2. Data were available in ten of the twelve cases including our case (3, 4, 7-13). Six of the patients were male and the age at the diagnosis ranged from 27 to 82 years (mean 51,3). Our case is the most elderly one among these cases. Tumor size varied from 4 to 28 cm (mean 12,2 cm). Tumors were located in lower pole of the kidney in four cases, upper pole in one case, entire kidney in one case and middle pole in three cases except the case reported by Nisimoto K in 1996. Our case is the second one located in renal sinus.

Histopathologic appearance of renal myxomas resembles others located at different sites of the body. The present case showed similar cellular structure with myxoma with the lack of pleomorphism, mitosis and nucleoli.

Renal myxoma should be differentiated from other benign and malignant mesenchymal tumors such as myxoid neurofibroma, myxoid leiomyoma, perineuroma, myxolipoma, myxoid variant malignant fibrous histiocytoma, liposarcoma, leiomyosarcoma and rhabdomyosarcoma. Renal myxoma can be easily distinguished from sarcoma with the lack of anaplastic features in nephrectomy material. Each benign tumor display characteristic histopathological and immunohistochemistry features (8, 11, 12). Our case did not have different histological areas such as neural, fibrous, leiomyomatous changes.

The present case was focally stained positive with SMA which reminded leiomyoma possibility in the differential diagnosis. However, specimens which were taken from the tumor did not show any fasciculated spindle cells or bundles that were specific for leiomyoma. Homogeneous structure throughout the sections, focal positive staining with SMA, desmin negativity and loss of parallel alignment of the cells also withdrew us from the diagnosis of leiomyoma (16). Our case also differed from myxoid liposarcoma with the lack of lipoblastic differentiation and differed from myxoid lipoblastoma with the lack of atypia, characteristic multilobular pattern and existence in the elderly (17, 18).

Cytopathological examination can be performed to distinguish benign tumors from malignancies. We have performed fine needle aspiration accompanied with CT before the surgical procedure and the result showed malignancy potential.

In conclusion, we described here a very rare type of mesenchymal tumor of the renal sinus. It is important to distinguish this benign entity from malignant tumors that may exhibit secondary myxoid changes due to overtreatment. The treatment of choice is radical nephrectomy. A case cured with enucleation was also reported in the literature but this was not possible in our case due to infiltration of the surrounding renal parenchyma.

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