Case Report Olgu Sunumu

Renal myxoma: a case report and review of the literature

Renal miksoma: Olgu sunumu ve literatürün gözden geçirilmesi

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Abstract

Myxomas are uncommon soft-tissue neoplasms, which are extremely rare in the kidney. In the present case, a right nephrectomy was performed because of suspected presence of a malignant renal tumor. In the macroscopic examination, the kidney contained a well-circumscribed, yellowish, and gelatinous tumor. The tumor was measured 6 cm in diameter. It was composed of fibroblast-like, slender spindle cells with myxoid stroma. With these histopathological findings, the case was diagnosed as renal myxoma. The case was discussed in the light of literature.

Key words: Kidney; myxoma; neoplasm.

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Özet

Miksoma nadir olarak görülen yumuşak doku tümörü olup, böbreklerde çok ender görülmektedir. Sunulan olguya şüpheli malign renal kitle nedeniyle sağ nefrektomi operasyonu uygulandı. Kitlenin makrosopik incelemesinde tümör iyi sınırlı, sarımsı ve jelatinöz bir yapıya sahipti. Tümör çapı en geniş yerinde 6 cm idi. Mikroskopik incelemede ise, fibroblast benzeri slender spindle hücreleri ile birlikte miksoid bir stroma gözlendi. Bu histopatolojik bulgular ile renal miksoma tanısı kondu. Olgu literatür eşliğinde tartışıldı.

Anahtar sözcükler: Böbrek; miksoma; neoplazi.

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Myxomas are extremely rare soft-tissue neoplasms that may be found in the kidney. The tumor is well-circumscribed, and it is composed of spindle shaped cells which showed diffuse immunoreactivity for vimentin, and reactivity was negative for S-100 protein, epithelial membran antigen, pancytokeratin, and smooth-muscle actin. Only ten cases have been reported in the literature. Here, we report an additional case treated with nephrectomy because of suspected presence of a malignant renal tumor.

Case report

A 59-year-old male was admitted to our hospital with a lower urinary tract symptoms. Ultrasonography of the abdomen demonstrated a well-defined mass occupying the upper portion of the right kidney. The computed tomography (CT) scan showed a 6 cm low-density tumor in the upper portion of the right kidney (Fig. 1). The routine chemistry and the hematologic assessments yielded normal results. A right nephrec-

tomy was performed. The nephrectomy specimen weighed 300 g except the perirenal fat. On sagittal section, a 6x5x4 cm tumor, which was easily distinguished from the adjacent renal parenchyma, was found. The mass was gray-white, semitranslucent, and gelatinous. Mucinous liquid was dripped from the cut surface (Fig. 2). No calculi were identified within the renal pelvis and ureter.

Histopathologic examination revealed a paucicellular and hypovascular tumor consisting of basophilic interstitial mucoid material containing sparse slender, oval, spindle, and stellate cells (Fig. 3). The myxoid material was stained positively with alcian blue and colloidal iron. Lipoblast, giant cells, mitoses, and necrosis were absent. Surrounding renal parenchyma was atrophic.

Immunohistochemicaly, the tumor cells were stained positively as diffuse for vimentin. Negative reactivity was for S-100 protein, epithelial membran antigen, pancytokeratin, and smooth-muscle actin. With these macroscopic, microscopic, and immuno-



Figure 1 Computer tomography scan showed a low-density, well-defined solid mass occupying the upper portion of the right kidney.

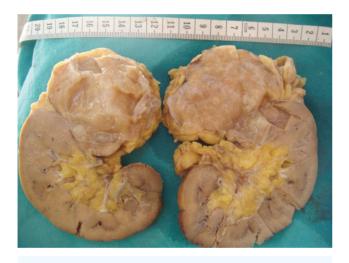


Figure 2 The upper pole of the right kidney contains a well demarcated, yellowish, and gelatinous tumor compressing the surrounding renal parenchyma and pelvis.

histochemical findings, the final histopathological diagnosis was primary renal myxoma.

Discussion

Myxomas were originally described by Virchow in 1863 with structures similar to the mucous tissue of the umblical cord. Myxomas can be identified in various anatomic locations and the currently recognized subtypes are restricted to the skin, soft tissues, bone, juxta-articular space, sinonasal cavity, maxillary antrum, and in organs or viscera. Organ myxomas may be observed in eye, heart, ovary, and kidney. [2-9]

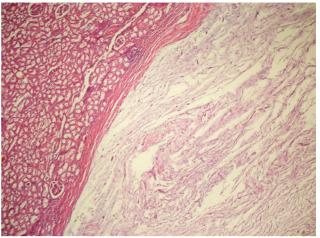


Figure 3 The microscopic examination showed the typical apperance of the myxoma that consisted of abundant myxoid stroma containing scattering fibroblast-like, slender spindle cells (H&E x40).

Primary true intrarenal myxomas are extremely rare. Until 1994, seven case reports of renal myxoma were published. In 1994, Melamed et al.^[3] presented two cases of renal myxoma and reinvestigated seven cases previously reported (including his two cases) as renal myxoma, and assumed that only three of them were pure renal myxomas. The remaining five cases exhibited features of sarcoma, fibroepithelial polip, or myxolipoma. They stressed the importance of distinguishing this benign tumor from other benign or malignant mesenchymal tumors.^[3] There have been three additional case reports on this entity recently. ^[2,7,9] Our case might be the eleventh case among the renal myxoma cases reported so far.

Age and gender in all renal myxoma cases were variable. Patients' ages range from 27 to 68 years (mean 47.5 years). The tumors are found more commonly in renal pole, and they vary in size from 4 to 28 cm (mean 10.5 cm) (Table 1).

The radiological appearance of the renal myxoma is various in CT and magnetic resonance imaging (MRI) investigations. In previous reported cases, it was found out that although CT and MRI of the abdomen demonstrated a well-defined mass occupying the middle of the right kidney, and a 8 cm low density solid tumor in the lower pole of the left kidney, in another study CT and MRI of the abdomen demonstated a well-defined semisolid/semicystic mass occupying the lower calyces of the left kidney.^[2,7,9] The findings of the first two studies share similarities

Reference	Age (year)/ gender	Presenting symptom	Side/size (cm)	Situation	Treatment
Bolat et al.[2]	27/female	Asymptomatic	Left/15	Lower pole	Nephrectomy
Melamed et al.[3]	52/female	Right renal colic	Left/7	Lower pole	Nephrectom
Melamed et al.[3]	68/female	Asymptomatic	Right/10	Upper pole	Nephrectom
Appel and Schoenberg ^[4]	NS	Hematuria for 2 months	Right/8	Parapelvic	Enucleation of mass
Kundu et al. ^[5]	36/male	Hypochondrium mass for 2 months	Left/28	Most of renal paranchyma	Nephrectom
Shenasky and Gillenwater ^[6]	62/male	Hematuria for 2 months	Right/4	Lower pole	Nephrectom
Owari et al.[7]	62/male	Asymptomatic	Right/8	Middle pole	Nephrectom
Val-Bernal et al.[8]	37/male	Asymptomatic	Right/6	Capsule	Nephrectom
Nishimoto et al.[9]	36/male	Asymptomatic	Left/9	Lower pole	Nephrectom
Present study	59/male	Lower urinary tract infection	Right/6	Upper pole	Nephrectom

wth our CT result which showed a 6 cm low density solid tumor in the upper pole of the right kidney (Fig. 1). However, in the last study, the appearance of the mass was different from our case in that it has semisolid/semicystic appearance in imaging methods. In the light of these data from literature, it can be concluded that renal myxoma has not typical CT and MRI appearance.

On sections, these tumors can easily be enucleated from the surrounding parenchyma. On histological examination, it was a tumor composed of fibroblastlike spindle cells and abundant myxoid stroma. No giant cells, mitoses, or necrosis were present. Immunohistochemically, the tumor cells were stained positive for vimentin, but negative for S-100 protein, epithelial membran antigen, pancytokeratin, and smooth-muscle actin. In ultrastructural examination, spindle-shaped, fibroblast like cells with long thin cytoplasmic processes, abundant and often dilated rough endoplasmic reticulum, a well-developed golgi complex, and secretory vesicles were disclosed. The nuclei were found to be oval or elongated, and a few with irregular nucleoli. There found to be no basal lamina around the tumor cells. These cells were extensively distinguished by abundant myxoid matrix containing scattered collagen fibrils.[8]

It is essential to distinguish renal myxoma from other tumors exhibiting prominent secondary myxoid feature. These are composed of perineurioma, myxoid neurofibroma, myxoid leiomyoma, myxolipoma, and myxoid variants of malignant fibrous histiocytoma, leiomyosarcoma, rhabdomyosarcoma, and extraskeletal chondrosarcoma. Grossly, all these neoplasm have an incostintent gelatinous property. However, each tumor has completely different ultrastructural, immunophenotypic, and genotypic features. In the particular case of renal sarcomas, increased cellularity, pleomorphism, high mitoses, well-developed vascularity, and necrosis are usually apparent. [3,7,8]

It has been reported that the patient with renal myxomas has no invasion, no metastasis, or no recurrence. As was in our case, the removal of the affected kidney was conducted in ten patients. Only in one patient with renal sinus myxoma, enucleation of the tumor was applied.^[4]

In conclusion, renal myxoma is a benign mesenchymal tumor. Since this tumor has a good prognosis, simple tumor enucleation might be a preferable option for the treatment instead of the complete removal of the kidney. It is essential to distinguish renal myxoma from other highly malignant neoplasms with secondary myxoid features.

Conflict of interest

No conflict of interest was declared by the authors.

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