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Primary renal myxoma: A case report

Primer renal miksom: Olgu sunumu

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ABSTRACT

Myxomas are uncommon soft-tissue neoplasms, which are extremely rare in the kidney with ten cases reported in the literature.

A 27-year-old woman was admitted to our hospital with epigastric pain and pelvic mass. Magnetic resonance imaging (MRI) of the abdomen showed a well defined 15x14x7 cm tumoral mass in the left kidney. The patient underwent a left-sided nephrectomy. On microscopic examination, the tumor consisted of large amounts of myxoid material containing a few uniformly scattered spindle cells. Immunohistochemically, tumor cells stained positive for vimentin, but not for S-100 protein, epithelial membrane antigen, pancytokeratin and smooth-muscle actin. With these histopathological and immunohistochemical findings, the case was diagnosed as 'renal myxoma'.

In this report, the clinical and histopathological findings, differential diagnosis and possible histogenesis of a case of myxoma that was originated from the left kidney of a 27 year-old female patient is presented.

Key words: Kidney, myxoma

ÖZET

Miksomlar nadir rastlanılan yumuşak doku tümörleridir, böbrekte oldukça ender görülür ve bugüne kadar literatürde yaklaşık on vaka bildirilmiştir.

Yirmi yedi yaşındaki kadın hasta epigastrik ağrı ve pelvik kitle nedeniyle hastanemize başvurmuştur. Batının manyetik rezonans görüntülemesinde (MRG) sol böbrekte 15x14x7 cm boyutlarında kitle saptandı. Hastaya sol nefrektomi yapıldı. Mikroskopik incelemede tümor, geniş miktarda miksoid materyal içinde dağınık az sayıda iğsi hücrelerden oluşmaktaydı. İmmünhistokimyasal olarak tümör hücrelerinde vimentin pozitif, S-100 protein, epitelyal membran antijen, pansitokeratin ve düz kas aktini negatif bulundu. Bu histopatolojik ve immünhistokimyasal bulgularla olguya 'renal miksom' tanısı konuldu.

Bu yazıda sol böbrekte yerleşen ender görülen miksom tanısı alan 27 yaşında kadın hastanın klinik ve histopatolojik bulguları, ayırıcı tanısı ve histogenezi sunulmaktadır.

Anahtar sözcükler: Böbrek, miksoma

INTRODUCTION

Myxomas may occur in a variety of tissues but are most common in skeletal muscle and generally follow an indolent course. Ten cases of kidney myxomas have been reported in the literature (1-5). The histogenesis of myxoma is still debated. Cardiac myxomas are thought to derive from multipotential mesenchymal cells, whereas their soft tissue counterpart is thought to originate from the primitive mesenchymal cells with fibroblastic features (1,3,4,6).

The differential diagnosis of myxoid tumors of the kidney includes many other benign and malignant soft-tissue lesions that exhibit prominent secondary myxoid features. It is important to recognize the existence of a renal myxoma to avoid confusing it with the malignant neoplasms having secondary myxoid features that may involve the kidney (1,3,4,6).

In this paper, the morphologic characteristics of this tumor are compared with other myxoid lesions of the kidney, which might cause confusion in differential diagnosis.

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CASE REPORT

A 27-year-old woman was admitted to our hospital with epigastric pain and pelvic mass incidentally diagnosed by ultrasonography during annual medical check-ups. The results of her physical examination were unremarkable, and laboratory values were within normal limits. Magnetic resonance imaging (MRI) of the abdomen demonstrated a well defined semisolid /semicystic mass occupying the lower calyces of the left kidney (Figure 1). On the MRI, the tumor was depicted as a homogeneous low-signal intensity on the T1 and heterogeneous high signal intensity on the T2-weighted pulse sequences.

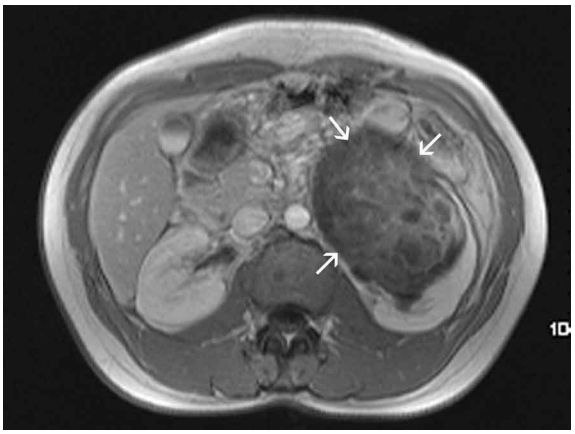

Figure 1. Magnetic resonance images demonstrate a tumor with heterogeneous high-signal intensity on the T2 - weighted pulse sequence (arrows).

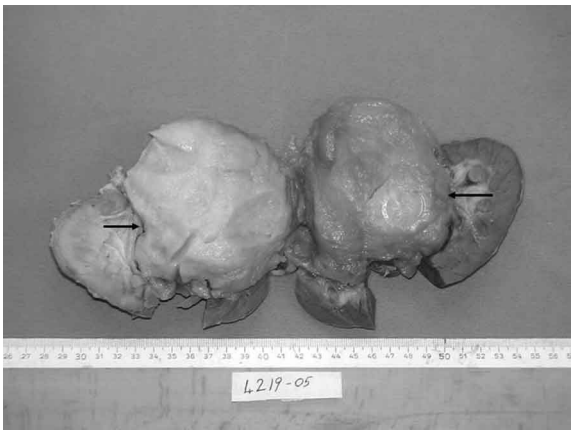

Figure 2. The middle pole of the left kidney contains a well-demarcated, yellowish, and gelatinous tumor compressing the surrounding renal parenchyma (arrows).

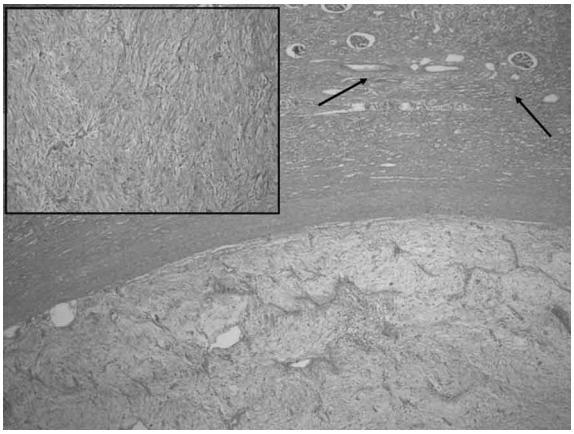

Figure 3. A fibrous pseudocapsule separates the sparsely cellular myxoid tissue from the renal parenchyma (arrows) (HE x100). Inset: Tumor showing spindled to polygonal cells in abundant myxoid stroma (HE x200).

The patient underwent a left-sided nephrectomy. The nephrectomy specimen weighed 14 gr excepting the perirenal fat. On serial sections, the lower pole of the left kidney demonstrated a 15x14x7 cm tumor surrounded by a thick, pearly white capsule which separated it from the adjacent renal parenchyma. The surface of the gray-white mass had a semitranslucent appearance and the mass was composed of a gelatinous, stringly mucoid material (Figure 2). No calculi were identified within the renal pelvis or proximal ureter. The surrounding kidney parenchyma appeared grossly unremarkable.

Histopathologic examination revealed a paucicellular and hypovascular tumor consisting of large amounts of basophilic interstitial mucoid material containing sparse slender, spindle, oval and stellate cells (Figure 3), and occasional fine strands of fibrous tissue. The myxoid material stained positively with Alcian blue. Nucleoli and mitotic figures were not evident.

No lipoblast, giant cells, or other spindle cell components were identified. The tissue at the interface between the myxoid stroma and the renal parenchyma showed thin bands of fibrous tissue forming a pseudocapsule. No obvious infiltration into the parenchyma was seen.

Immunohistochemically, tumor cells were stained positively with vimentin, while S100

protein, epithelial membrane antigen (EMA), pancytokeratin and smooth-muscle actin (SMA) did not show any positive reaction. With the above mentioned gross, microscopic and immunohistochemical findings, the final histopathological diagnosis was myxoma of the kidney. The patient is free of disease for one year.

DISCUSSION

Myxomas were originally described by Virchow in 1863 (7), and Stout (8) established the basic histologic criteria for diagnosis in 1948. Enzinger, in 1965, suggested that the lack of aggressive growth and the paucity of vascular structures suggested a similarity to fibroblasts and a close relationship to the early developmental stage of ganglia (9).

The histogenesis of renal myxoma remains unresolved and speculative. Some authors have considered it to be a degenerative change seen in adipose tissue in brown atrophy of the heart or similar to the myxoid changes in uterine leiomyomas. The others believe that the uniform cellular component throughout the lesion supports a neoplastic nature rather than a regressive change within a preexisting tumor. The cellular origins of the tumor have been contemplated to be primitive mesenchymal cells with fibroblast like features (1).

Myxomas can be located in skin, soft tissues, bone, juxta-articular space, sinonasal cavity, maxillary antrum, and in various organs or viscera (10). Myxomas may be observed in eye (11), heart (12), ovary (13), and kidney (1-5).

Between 1887 and 1995 nine case reports of renal myxoma were published. Melamed et al. reviewed these nine cases, and assumed that only three of them were pure renal myxomas (incl. their two cases) (1). There have been two additional case reports on this entity within the last year (4, 5). Our case might be the sixth case of pure renal myxoma.

Intrarenal myxomas have no gender predilection. Patients' ages range from 36-68 years (mean 54.5 years). The lesions are found more commonly within renal poles and they vary in size from 4 to 28 cm (mean 12.2 cm) (6). They have a smooth capsule and a solid-cystic cut surface. This appearance is not specific for the renal myxoma. Histologic features are distinctive, and our case resembles to the other renal myxomas that were previously described in the literature

Renal myxoma is a very rare tumor composed of fibroblast-like spindle cells and abundant myxoid stroma. It is essential to distinguish renal myxoma from malignant and benign mesenchymal tumors exhibiting prominent secondary myxoid features. The spectrum embraces perineurioma, myxoid neurofibroma, myxoid leiomyoma, myxolipoma, low grade fibromyxoid sarcoma, the myxoid variants of malignant fibrous histiocytoma (low grade myxofibrosarcoma), liposarcoma, leiomyosarcoma, rhabdomyosarcoma, and extraskeletal chondrosarcoma. Grossly, all these neoplasms have a variable gelatinous consistency. However, each tumor has a complete set of exclusive morphologic, ultrastructural, immunophenotypic, and genotyphic features. In the particular case of renal sarcomas, increased cellularity, pleomorphism, mitoses, well-developed vascularity, and necrosis are usually apparent (1,4,6).

Myxoid stroma has also been found in sarcomatoid renal cell carcinoma, as well as in the sarcomatoid variant of transitional cell carcinoma (14,15). These changes are usually focal, and an appropriate sampling will demonstrate adjacent anaplastic spindle cells together with areas of epithelial differentiation.

Renomedullary interstitial cell tumors are the commonest benign mesencyhmal lesions of the kidney (16). Occasionally they may be highly myxoid but usually have a dense fibrous stroma with areas of hyalinization. Rarely do they grow to large size, however, because of their uniform cell population they are considered neoplasms rather than hamartomas (1).

In conclusion, we have described an unu-

sual case of renal myxoma. It is important to distinguish this benign tumor from many other highly malignant neoplasms that may involve the kidney and exhibit secondary myxoid features. Myxomas have an excellent clinical outcome after radical excision and their recognition is essential to avoid an overtreatment .

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