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Renal myxoma: a case report

Mixoma renal: relato de caso

Carlos Henrique C Souza[1]; Kennedy S. Carneiro[2]; Katia R. M. Leite[3]; Alcendino A. Junior[4]; Fredwilson S. Costa[1]

1. Medical Student. 2. Universidade Federal de São Paulo (UNIFESP); Faculdade de Medicina da Universidade Severino Sombra (USS), Vassouras-RJ. 3. UNIFESP; Faculdade de Medicina da Universidade de São Paulo (FMUSP). 4. Universidade do Estado do Rio de Janeiro (UERJ); Faculdade de Medicina da Universidade Severino Sombra, Vassouras-RJ.

ABSTRACT

Myxomas are rare tumors that can appear in many anatomical locations. There are only 14 cases of renal involvement documented in the literature. This article reports a case of renal myxoma in an elderly woman with recurrent cystitis. After five years of follow-up, the computed tomography (CT) revealed a large solid tumor mass in the left kidney. Tumor resection was performed preserving the affected kidney with histopathological diagnosis of renal myxoma. The objective of this study is to report a rare case of renal myxoma, emphasizing the importance of the differential diagnosis from other benign and malignant mesenchymal tumors.

Key words: renal myxoma; kidney; myxoma; renal neoplasm.

INTRODUCTION

Myxomas are uncommon benign neoplasms, extremely rare in the kidney. They can be found in the skin, soft tissues, bones, joint spaces, paranasal sinuses, maxillary antrum, viscera[1], and intramuscular[2, 3]. The affected viscera are eyes[4], heart[5], ovaries[6] and kidneys[2, 3, 7-15].

Only 14 cases of patients with renal myxomas have been reported in the literature[3, 16], none preceded by renal cyst. The tumor is well-circumscribed and is composed of spindle-shaped cells, scattered in a myxoid stroma, showing immunoreactivity for vimentin and negative reactivity for S-100 protein, epithelial membrane antigen (EMA), pancytokeratin and smooth muscle actin[2, 7-15]. There is still no description of this type of renal tumor in the literature. The importance of the recognition of its existence is to prevent diagnostic errors with malignant and benign neoplasms that present secondary myxoid features, which may involve the kidney. This paper reports the second case of renal myxoma in which was performed tumor excision only, and the affected kidney was preserved.

CASE REPORT

The patient was a white woman, aged 73 years, showing recurrent cystitis since 2003. In 2005, after new bouts of cystitis, an ultrasonography (US) was performed and bilateral renal cysts were detected, they were 4.2 cm in the left kidney and 1.4 cm in the right kidney. In 2007, she complained of right-sided cramp-like lumbar pain, irradiating anteriorly, with nausea, negative renal fist percussion, and right-sided flank pain at palpation. Computed tomography (CT) was ordered, and showed kidneys with reduced volume, lobulated contour, preserved parenchymal thickness, bilateral renal cortical with rounded and hypodenses formations, (cysts) without significant enhance after contrast medium, measuring 7.4 × 6.8 cm in the middle third of the left kidney (**Figure 1**). After six months, the control CT scanning showed, besides the cortical cysts, the presence of a solid tumor mass, enhanced by contrast medium, measuring 11.9 × 10.1 cm, with well-defined edges in the left flank, suggesting neoplasia (**Figure 2**). There were no other changes in biochemical and urinalysis tests.

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The patient underwent partial nephrectomy due to the exophytic tumor found with gelatinous and well-defined aspect, with narrow pedicle, without lymphadenopathy. The histopathologic diagnosis was myxoma. It showed short spindle-shaped cells with small, oval, delicate nuclei, without hyperchromasia or mitotic activity. Cytoplasm was reticular diffuse and eosinophilic. The cells were involved in a myxoid stroma highly vascularized. Immunohistochemistry showed unique positive tumor cells for vimentin. However, the de S-100 protein, smooth muscle actin, cytokeratin, and murine double minute 2 (MDM2) expression were negative (**Figures 3** and **4**).

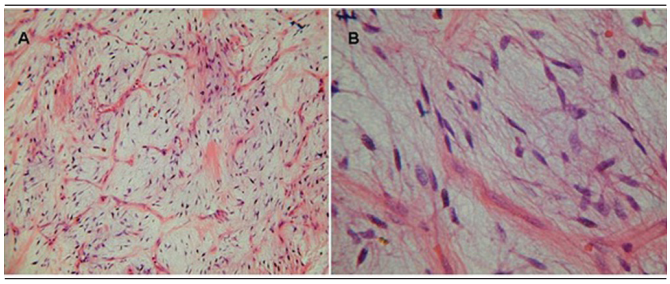

FIGURE 3 – *Benign mesenchymal neoplasm consists of scattered spindle cells in myxoid stroma. No mitotic activity or pleomorphism is observed*
3A): HE, 100×; 3B) HE, 400×.
HE: hematoxylin and eosin.

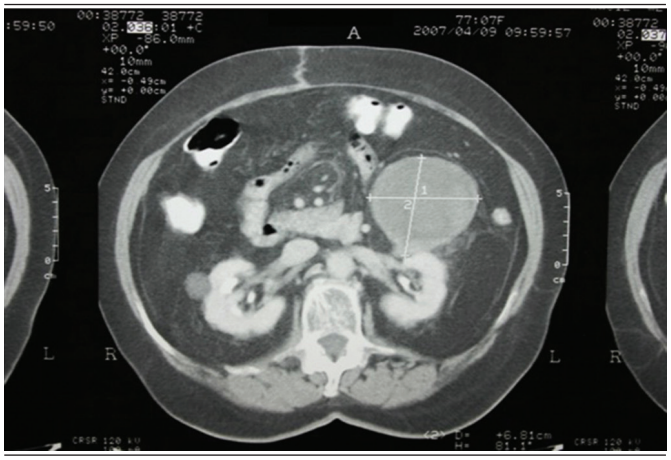

FIGURE 1 – *Kidneys normally located, reduced volume, lobulated contour, cortical parenchymal thickness preserved with rounded and hypodenses formations without significant enhancement after contrast medium, measuring 7.4 × 6.8 cm the largest cm in the middle third of the left kidney*

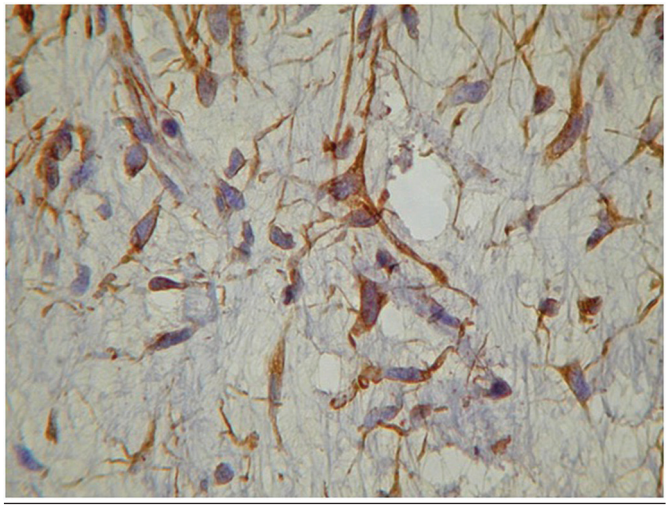

FIGURE 4 – *Immunohistochemistrystudy showing only positive tumor cells for vimentin (400×)*

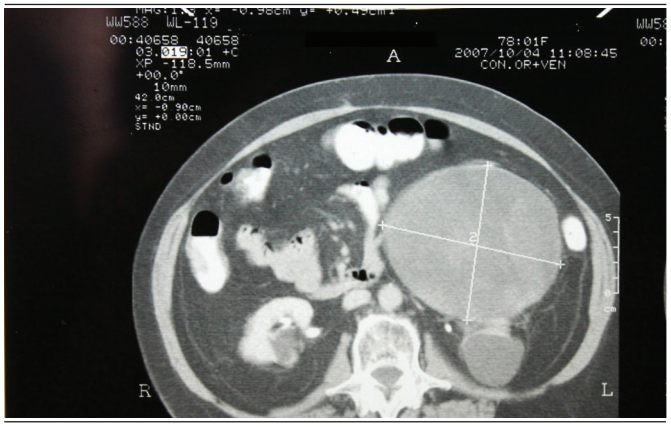

FIGURE 2 – *Presence of large tumor mass, solid, enhanced by iodinated contrast medium, not calcified, irregular contour, projected on the left flank, anteromedial and exophytic flank in relation to the left kidney, measuring 119.7 × 101.9 mm, suggesting renal neoplasm. Identification of renal cortical cysts*

DISCUSSION

It was Virchow[17] who first introduced the term myxoma, and Stout[18] established that the myxoma is a tumor composed of stellate or spindle-shaped cells, with myxoid stroma containing mucopolysaccharide, through which delicate reticular fibers run in various directions. We also examined and determined criteria for distinguishing myxomas from sarcoma variations and mesenchymal neoplasms.

Currently, there is no records of specific clinical presentation for renal myxoma[16] and, due to its rarity, renal myxoma is often confused with other malignant lesions[16]. It is known so far that there is no invasion, metastasis or recurrence[8]. In our

renal myxoma case, the patient had concomitant kidney cysts, and in a recent article published, there was also, hemorrhagic cysts concomitant with myxoma[16]. Its differential diagnosis includes a variety of benign and malignant mesenchymal tumors that may show occasionally prominent myxoid features[2], namely: perineurioma, myxoid neurofibroma, myxoid leiomyoma, myxolipoma and myxoid variant of malignant fibrous histiocytoma, leiomyosarcoma[19], rhabdomyosarcoma and extraskeletal chondrosarcoma[3, 7, 8], low-grade fibromyxoid sarcoma[20, 21], and solitary fibrous tumor[22]. There is still no description of renal myxomas in the literature[3].

Macroscopically and microscopically, renal myxomas resemble the primitive mesenchyme. The tumor may be found in different locations in the body, the most common is intramuscular[2, 3]. It is believed that it is originated in fibroblasts that have lost the ability to polymerize collagen[3], however, there are controversies in the literature[2, 10, 15]. 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[2, 10, 15].

Macroscopically, the tumor has gelatinous aspect and is well-defined[8]. Histopathologically, it is composed of thin fibroblasts-like spindle-shaped cells, scattered in an abundant myxoid stroma, closely resembling primitive mesenchyme and myxomas of other sites in the body[3]. It is considered a benign fibroblastic tumor because mitotic activity and cell pleomorphism are not present[8].

In a wide literature review in 1994, Melamed *et al*. assert that only five case reports were truly renal myxomas, including their two cases. The remaining cases exhibit features of sarcoma, fibroepithelial, polyp or myxolipoma[2].

The immunohistochemical findings of previous cases of renal myxoma, tumor cells stained positive for vimentin, but negative for S-100 protein, EMA, pancytokeratin and smooth muscle actin[2, 7-15], except in one case, that was focally positive for smooth muscle actin[3].

The imaging exams demonstrate the renal myxoma as a large heterogeneous mass, predominantly hyperechoic on US and hypodense on CT scan, with more homogeneous signal on magnetic resonance imaging (MRI), which presents with low signal intensity on T1 and hyperintense on T2. Its contours are relatively regular, discreetly multilobulated, and its interface is well-defined with the adjacent renal parenchyma, only shifting the structures without invading them[23].

Radical nephrectomy is considered the treatment of choice, showing no recurrence or metastasis in any case[8]. In one case only tumor enucleation was performed[24]. Our case is the second in which there was kidney preservation, and the second that had renal cysts. As it is a benign tumor, we agree that, under the guidance of imaging examination, percutaneous biopsy may be a better option in the future for renal preservation, enabling better operational planning with maximum preservation of the affected kidney[16].

The patient is currently being monitored, free of recurrence and metastasis.

In conclusion, renal myxoma is a rare tumor with good prognosis and has its origin in fibroblasts[3]. The differential diagnosis is important to avoid confusing it with a variety of malignant and benign mesenchymal tumors such as sarcomatoid carcinoma, which may show secondary myxoid features[2]. The best treatment option would be, when conditions are favorable, tumor enucleation with the preservation of the affected kidney.

RESUMO

Mixomas são tumores raros que podem ser encontrados em muitas localizações anatômicas. Na literatura, há apenas 14 casos de acometimento renal. Neste artigo, é relatado um caso de mixoma renal em mulher idosa com cistites de repetição. Após cinco anos de acompanhamento, a tomografia computadorizada (TC) evidenciou grande massa tumoral sólida em rim esquerdo. Realizou-se exérese do tumor preservando o restante do rim afetado com diagnóstico histopatológico de mixoma renal. O objetivo deste trabalho é relatar um caso raro de mixoma renal, enfatizando a importância do diagnóstico diferencial de outros tumores mesenquimais benignos e malignos.

Unitermos: *mixoma renal; rim; mixoma; neoplasia renal.*

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MAILING ADDRESS

Carlos Henrique Câmara de Souza

Universidade Severino Sombra, Centro de Ciências da Saúde, Faculdade de Medicina; Av. Expedicionário Oswaldo de Almeida Ramos, 280; Centro; CEP: 27700-000; Vassouras-RJ, Brazil; e-mail: carloshenrique.camara@hotmail.com.