Myxoma versus myxoid liposarcoma involving the kidneys

Sarah K. Findeis, MD^a , Mauricio N. Salicru, MD^b , and Atin Agarwal, MD^a

^aDepartment of Pathology, Baylor University Medical Center, Dallas, Texas; ^bDepartment of Pathology, Methodist Dallas Medical Center, Dallas, Texas

ABSTRACT

Myxoid lesions of the kidney are rare. We present a case of a 74-year-old man who presented with an 8.5 × 8.0 × 6.0 cm left kidney mass that was grossly confined to the kidney and had a gelatinous cut surface. Histology of the tumor showed bland spindle cells in a myxoid stroma with interspersed thin-walled vessels. The tumor was negative for smooth muscle actin, desmin, CD34 (highlighted vessels), S100, and HMB-45 by immunohistochemistry. There was focal, nonspecific staining of MDM2 and CDK4. The lesion appeared more vascular than would be expected for a classic myxoma and, therefore, fluorescence in situ hybridization was performed for the 12q13 (*DDIT3* or *CHOP*) rearrangement to rule out myxoid liposarcoma and the result was negative for a rearrangement. This case highlights the difficulty of delineating a primary myxoma of the kidney from a well-differentiated myxoid liposarcoma.

KEYWORDS CHOP; DDIT3; myxoid mass; myxoma of the kidney; renal myxoma; well-differentiated myxoid liposarcoma

The kidney is an unusual site for a myxoid lesion. When approaching such a lesion, the differential diagnosis includes both benign and malignant soft tissue neoplasms as well as metastasis. After ruling out epithelial neoplasms, which is usually readily achieved by morphology and/or immunohistochemistry stains, the histologic approach to a primary myxoid lesion of the kidney involves identifying whether there is any epithelial component. For a lesion lacking an epithelial component, the differential narrows to myxoma, myxoid neurofibroma, perineuroma, myxoid leiomyoma, myxolipoma, and a sarcoma with a myxoid element (myxoid liposarcoma, low-grade myxofibrosarcoma, low-grade fibromyxoid sarcoma, leiomyosarcoma, rhabdomyosarcoma, extraskeletal chondrosarcoma).¹ All of these entities as kidney primaries are extremely rare and therefore immunohistochemical and molecular testing is required.

CASE REPORT

A 74-year-old man presented with bilateral renal masses with a history of clinical stage 4 prostate cancer and severe renal insufficiency (creatinine 6.42 mg/dL). Other pertinent past medical history included severe coronary artery disease,

postcoronary artery bypass. The masses were identified by the prostatic cancer staging computed tomography scan. The left kidney had two masses: one in the upper pole and the other in the posterior aspect. A right solid renal mass was also seen. The patient underwent bilateral nephrectomies.

One of the masses within the left kidney was an 8.5 × 8.0 × 6.0 cm midposterior exophytic mass involving the cortex with a red-tan, gelatinous cut surface. The tumor did not invade the capsule, renal sinus fat, or renal vein. The second left kidney mass was yellow and well circumscribed, which microscopically was an adrenal cortical adenoma. The solid right renal mass was not grossly identified; however, there were multiple simple corticomedullary cysts.

Microscopic examination showed a relatively well-circumscribed lesion with a thinned capsule. The lesion was located within the renal parenchyma (*Figure 1a*). The tumor was composed of bland fibroblastic-like spindle cells in a myxoid stroma with focal areas of hypercellularity without mitoses. Focally, there were few entrapped adipocytes, but lipoblasts were not seen. The lesion showed vessels arranged in a chicken-wire pattern (*Figures 1b, 1c*).

The tumor was negative for smooth muscle actin, desmin, S100, HMB-45, and CD34 (highlighted vessels). There was focal nonspecific staining for MDM2 and CDK4.

Corresponding author: Sarah K. Findeis, MD, Department of Pathology, Baylor University Medical Center, 3500 Gaston Ave., Dallas, TX 75246 (e-mail: Sarah.Findeis@bswhealth.org)

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Figure 1. (a) Tumor and adjacent kidney parenchyma. (b) Low-power view of tumor in relation to Gerota's fascia (inked black). (c) Chicken-wire vasculature of tumor with myxoid stroma and scattered inflammatory cells.

MUC4 was negative, ruling out low-grade fibromyxoid sarcoma.² Because of the increased vascularity and slightly increased cellularity, fluorescent in situ hybridization (FISH) was performed for the 12q13 (*DDIT3* or *CHOP*)

rearrangement (Mayo Laboratories), which came back negative (although a negative result could not fully exclude myxoid liposarcoma). Based on the immunohistochemistry and FISH studies, the diagnosis of myxoma was rendered.

DISCUSSION

After immunohistochemical stains ruled out melanoma, low-grade fibromyxoid sarcoma, and neural-derived lesions, our differential diagnosis was between myxoma and myxoid liposarcoma. To the best of our knowledge, primary myxomas of the kidney are limited to only 17 documented cases in the English literature. Primary myxoid liposarcomas are even more rare, with one confirmed case in the English literature that arose from the fatty renal capsule.^{3*} It should be noted that Carney syndrome was considered part of the differential diagnosis; however, the patient was older and had no history of other myxomas or skin hyperpigmentation.

Generally, myxomas are composed of eosinophilic stellate cells in a mucoid background with scattered thin-walled vessels and most frequently occur in the heart. Historically, lesions that are histologically similar to cardiac myxomas have been classified as myxomas, regardless of location.⁴ However, this is likely a misconception, because there are differences in clinical demographics, tumor behavior, and histology (although minor) depending on the site.⁴ Most cases of renal myxomas have been surgically resected via nephrectomy because the initial impression was a malignant renal mass.¹ Typically, excision is curative,⁵ with evidence that limiting excision to the tumor with preservation of kidney parenchyma is adequate treatment.⁶

The *DDIT3* gene break-apart has been shown to be a highly sensitive and specific assay,^{7,8} and all cases (100%) from a small study ($n = 5$) of retroperitoneal myxoid liposarcomas had the *DDIT3* break-apart.⁹ However, the laboratory where the FISH analysis was performed for our case reports a rearrangement of *DDIT3* (*CHOP*) in 18 of 26 (69%) myxoid/round cell liposarcoma specimens.¹⁰ In 2011, Narendra et al⁷ reported that there was a case of myxoid liposarcoma that was diagnosed based on morphology but was negative for *DDIT3* break-apart. However, in this study, the authors reasoned that because the case showed amplification of *DDIT3*, which was not seen in the 14 cases of established myxoid liposarcoma, it may not actually be a myxoid liposarcoma but instead another type of liposarcoma.

This case highlights the difficulty of diagnosing primary myxoid lesions of the kidney. Utilization of immunohistochemical stains and molecular testing is imperative in whittling down the differential diagnosis. Clinical correlation should be recommended, especially when there are atypical features.

*The article by Zhang et al³ reports a case of myxoid liposarcoma; however, the paper reviews two other cases of pure myxoid liposarcoma that are published in Chinese.

ORCID

Sarah K. Findeis <http://orcid.org/0000-0002-5833-0577>

Mauricio N. Salicru <http://orcid.org/0000-0002-1886-8081>

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