Solitary fibrous tumor of the renal hilum: A rare diagnosis for a not so rare clinical picture

Tumor fibroso solitario del hilio renal: un diagnóstico poco común de un cuadro clínico no tan raro

Dear Editor:

Imaging studies such as computed tomography (CT) or ultrasound are becoming more and more frequent in our routine clinical practice. Often these techniques identified masses in the kidney but cannot always distinguish between renal cell carcinomas, metastases, carcinomas of the upper urinary tract, or benign renal tumors.

Surgical resection, which provides tissue for accurate diagnosis as well as definitive therapy, remains the standard approach for most patients who present with renal masses. However, a common clinical problem is the management of patients with small lesions or those of uncertain malignant potential. Also, elderly patients with multiple comorbidities represent a subgroup in which a customized approach is frequently required.

Percutaneous biopsy or fine needle aspiration with image guidance may be an alternative in selected cases. Yet, these techniques have not been widely utilized, because of concern about haemorrhage or tumor seeding along the needle track. Nevertheless, percutaneous biopsy may be an alternative in selected patients in whom surgery may entail significant risk.

In this regard, we saw in our institution a 47-year-old female in whom an abdominal CT scan performed as a study of unspecific abdominal pain revealed an ovoid mass of 26 mm × 17 mm × 28 mm. compressing the right renal pelvis with avid contrast enhancement and intra-lesion calcifications and no signs of disease spread (Fig. 1a). A radical nephrectomy was performed and histopathological analysis showed a solitary fibrous tumor (SFT). The renal hilum contained a 2.3-cm mass of adipose tissue. Microscopically, the neoplasm was separated from the kidney and the renal pelvis parenchyma and it was composed of round to spindle-shaped tumour cells arranged in hypercellular and hypocellular areas separated by thick bands of collagen. No necrosis or atypia were observed and the mitotic index was low. Tumor cells stained positive for CD34, CD99, Vimentin Bcl2 and no staining was observed for AE1/AE3, MelanA, HMB45, smooth muscle actin, CD10, Desmin and S-100 protein (Fig. 1b).

SFT is an unusual mesenchymal neoplasm that shows a prominent hemangiopericytoma (HPC)-like branching vascular pattern, and in fact SFT and HPC share histological features and have almost identical gene expression profiles, so they are now grouped as one entity.

SFT/HPC is more common in middle-aged adults and it occurs most frequently in the pleura. Surgical resection is the gold standard and it is curative in many cases. Nevertheless, some reports reveal a metastatic potential even early in the course of the disease.

SFT/HPC is rarely diagnosed in genitourinary (GU) oncology. It presents as well circumscribed masses that histologically consist of tightly packed spindle cells separated by bands of hyalinized collagen and branching blood vessels. Most SFT/HPC are indolent although some can behave as high-grade sarcomas. There is a rare form (i.e. Doege-Potter syndrome) where patients present with very large masses and hypoglycemia induced by tumor cells production

Figure 1  (a) Abdominal computed tomography showing a pararenal mass with contrast enhancement. (b) Hematoxylin and eosin-stained paraffin section showing the SFT resected and its immunohistochemistry staining with positivity for CD99 and CD34.

of IGF-2. Radiological findings in the diagnosis of SFT usually reveal large tumors, with discrete margins and avid contrast enhancement. Resection of these tumors is curative in most cases, but rare recurrences might occur mostly in tumors larger than 10 cm. Doxorubicin-based chemotherapy has scarce activity in the metastatic or unresectable setting. Targeted therapy inhibiting platelet-derived growth factor receptor or vascular endothelial growth factor has recently shown clinical activity in these patients and can represent an alternative treatment.

This case illustrates a rare entity in GU oncology and attempts to summarize its clinical presentation, pathology, and management. It also shows how accurate diagnosis of renal masses remains a challenge for the physicians involved in GU oncology.

References


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