CLINICAL CASE

Forniceal rupture and urinoma secondary to retroperitoneal fibrosis: a clinical case and literature review

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KEYWORDS
Retroperitoneal fibrosis; Images; Hydronephrosis; Forniceal rupture; Computed tomography; Urinoma.

Abstract  The most frequent site of excretory system rupture that is secondary to obstruction, usually arising from stone disease, is the renal fornix. Forniceal rupture and the formation of retroperitoneal fluid collections are rare forms of retroperitoneal fibrosis. Presented herein is the case of a man in the sixth decade of life that came to the emergency service because of abdominal pain. Computed tomography scan of the abdomen and pelvis revealed a slightly enhanced retroperitoneal soft tissue lesion that extrinsically enveloped and compressed the right ureter. A second image of lower density was observed in the excretory phase that proved to be a urinoma secondary to rupture of the ipsilateral renal fornix. A double-J catheter was placed to decompress the excretory system and systemic steroid therapy was given, with good response. Retroperitoneal fibrosis is an uncommon disease characterized by the development of inflammation and fibrosis in the retroperitoneal space. We describe herein two forms, idiopathic and secondary, the former being more frequent. Current evidence has associated the idiopathic variant with IgG4-related diseases.

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PALABRAS CLAVES
Fibrosis retroperitoneal;

Hipogonadismo de inicio tardío: revisión de conceptos y pautas diagnósticas

Resumen  El sitio de rotura secundaria a obstrucción más frecuente del sistema excretor es el fórmix renal, habitualmente de origen litiásico. La rotura fornicial y la formación de colecciones líquidas retroperitoneales son formas de presentación muy infrecuentes de la fibrosis.

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Introduction
The most frequent excretory system site of rupture secondary to obstruction is the renal fornix, in accordance with Laplace’s law. Rupture is secondary to ureteral obstruction arising from lithiasis in 74% of the cases.1 Forniceal rupture and the formation of retroperitoneal fluid collections are rare forms of retroperitoneal fibrosis that require a high degree of diagnostic suspicion. Routine complementary studies with elimination phases are a necessity when excretory system rupture is suspected.

Clinical case
A 55-year-old man had a past history of diabetes mellitus, high blood pressure, and dyslipidemia, with moderate metabolic control. He sought medical attention due to nonspecific abdominal pain. An abdominopelvic computed tomography scan was ordered (fig. 1A-D) that identified tissue with soft tissue density (30 HU) and poorly defined contours. It surrounded the infrarenal artery and partially surrounded the common iliac arteries without significantly displacing them. Maximum contrast-enhancement reached 70 HU in the portal venous phase. An unenhanced low density zone stood out that extended from the right renal hilum into the caudal and perirenal space and was delimited by the psoas muscle, the inferior vena cava, and the duodenum. The impregnation of the entire right kidney was also delayed. The elimination phase confirmed the suspicion of excretory system rupture, characterized by contrast medium filtration adjacent to the fornix of the upper pole and extending into the previously described hypodense collection (figs. 2A and B). Moderate hydroureretonephrosis was conspicuous with traction of the ureter toward the midline and stricture in its middle third. A double-J catheter was placed to decompress the urinary tract and systemic steroid therapy was administered, with good treatment response.

Discussion
Retroperitoneal fibrosis is a rare disease characterized by the development of inflammation and fibrosis in the retroperitoneal space. It can extend from the crura of the diaphragm to the bifurcation of the iliac vessels, although it usually surrounds the infrarenal aorta, the inferior vena cava, the ureters, and the iliac vessels and can, albeit infrequently, affect the neighboring solid organs.2

One idiopathic and one secondary variant are described herein. Two-thirds of the cases correspond to the idiopathic form of the disease, which was initially considered an excessive inflammatory response to antigens derived from the low-density lipoprotein oxidation that is present in atherosclerotic plaques.3 Today, however, it is thought that atherosclerosis is only a predisposing condition in susceptible individuals and that idiopathic retroperitoneal fibrosis is a manifestation of a systemic inflammatory/autoimmune pathology in the context of IgG4-related diseases.2 The remaining 33% of the cases are secondary to other factors, mainly drugs (ergot alkaloid derivatives) and neoplasias (lymphoma, retroperitoneal sarcoma, carcinoids, and metastasis) at 12 and 10%, respectively.4

There are no imaging study findings that enable benign variants to be differentiated from malignant ones with precision. Nevertheless, reports state that displacement of structures, a nodular aspect, and being situated over the origin of the renal arteries are correlated with malignancy. And despite the apparent contradiction, an infiltrative aspect is suggestive of benign etiology, having a low intensity signal in heavily T2-weighted magnetic resonance sequences.5

Clinical symptoms are initially nonspecific, such as general malaise, anorexia, low weight, fever, and lumbar or flank pain. Inflammatory tissue surrounds and compresses one or both ureters in 56-100% of the affected patients, and can manifest as signs and symptoms of acute, as well as chronic, kidney failure. There can also be renovascular arterial hypertension and kidney failure as a consequence of renal vascular pedicle involvement.4 Extrinsic arterial, venous, and lymphatic compression can progress to claudication, thrombosis, venous congestion, and lower limb edema.5

Ethical responsibilities
Protection of persons and animals. The authors declare that no experiments were performed on humans or animals for this study.
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Data confidentiality. The authors declare that no patient data appear in this article.

Right to privacy and informed consent. The authors declare that no patient data appear in this article.

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Conflict of interest

The authors declare that there is no conflict of interest.

References


Figure 1  A. Non-contrast-enhanced CT. A low-density zone is observed that extends from the right renal hilum to the perirenal space. B. Contrast-enhanced CT in the portal venous phase. The same level as A, showing the absence of enhancement in the previously described hypodense zone, thickening of the urothelium, and delay in the impregnation of the entire right kidney. C. Non-contrast-enhanced CT. More caudal than A, showing density in the soft tissue with poorly defined contours, surrounding the infrarenal aorta that is not displaced. The caudal extension of the hypodense zone is also identified. D. Contrast-enhanced CT in the portal venous phase. The same level as B. There is mild enhancement of the peri-aortic tissue, with no impregnation of the hypodense zone.
Figure 2A.  *Oblique multiplanar CT reconstruction in the elimination phase.* Contrast medium filtration is shown, adjacent to a renal fornix of the upper pole extending into the previously described hypodense collection. B.  *Volumetric reconstruction.* Moderate right hydroureteronephrosis is identified with traction of the ureter to the midline and stricture in its middle third portion. There is a collection adjacent to the ureteral trajectory.